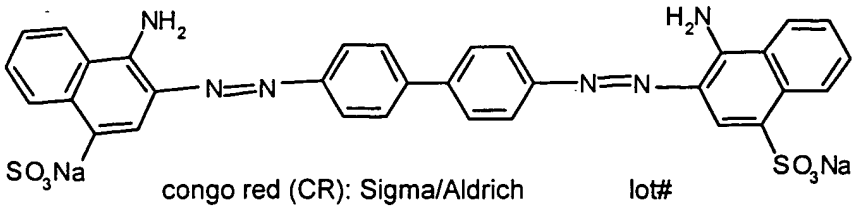
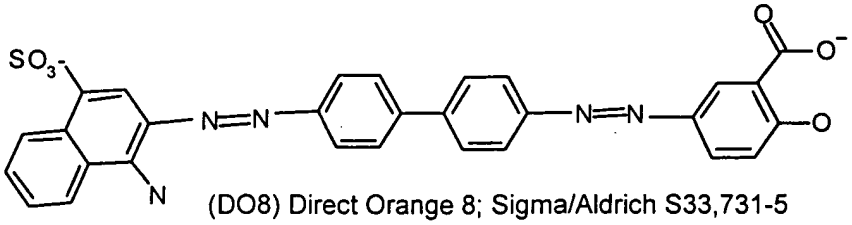
Fig.1A



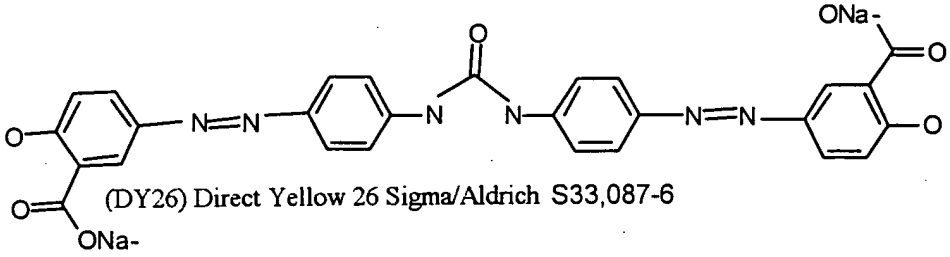
congo red (CR): Sigma/Aldrich lot#

Fig.1B



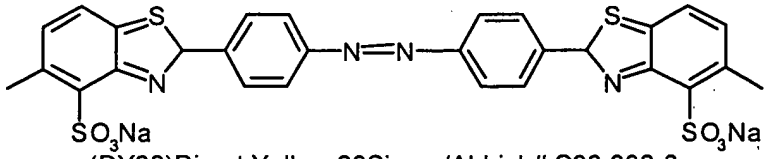
(DO8) Direct Orange 8; Sigma/Aldrich S33,731-5

Fig.1C



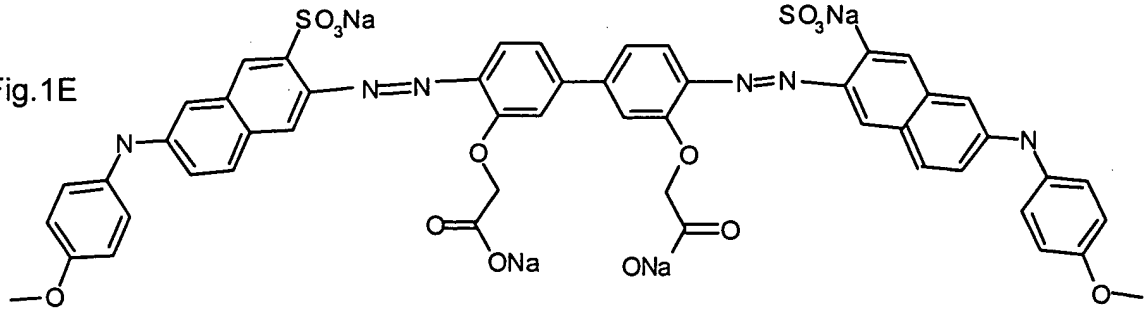
(DY26) Direct Yellow 26 Sigma/Aldrich S33,087-6

Fig.1D



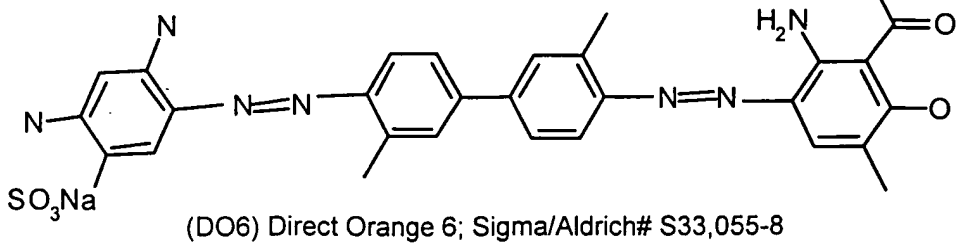
(DY28)Direct Yellow 28Sigma/Aldrich# S33,038-8

Fig.1E



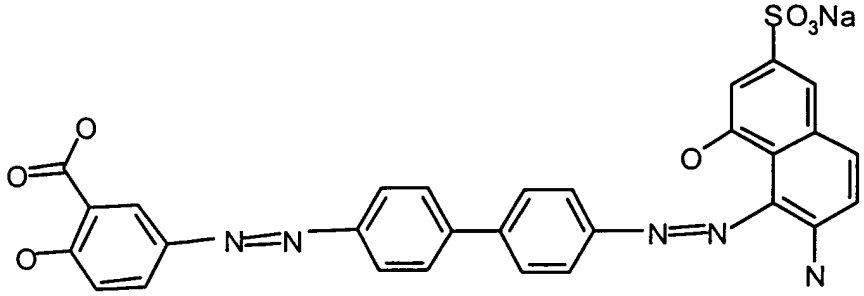
(DB158) Direct Blue 158; Sigma/Aldrich# S32,721-2

Fig.1F



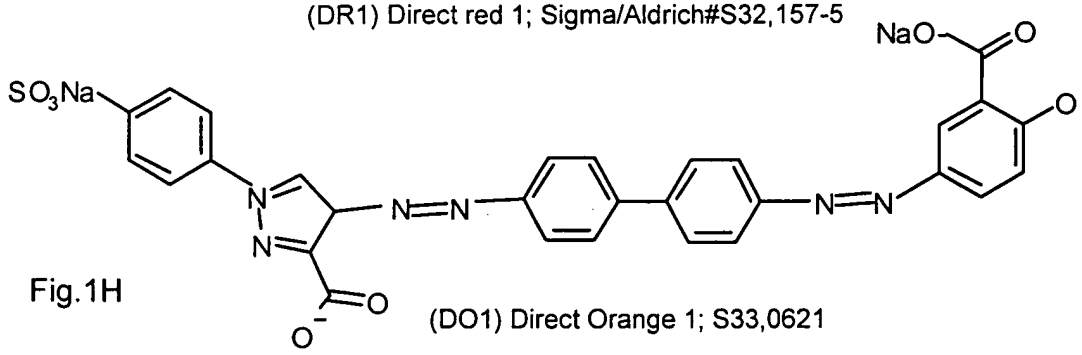
(DO6) Direct Orange 6; Sigma/Aldrich# S33,055-8

Fig.1G



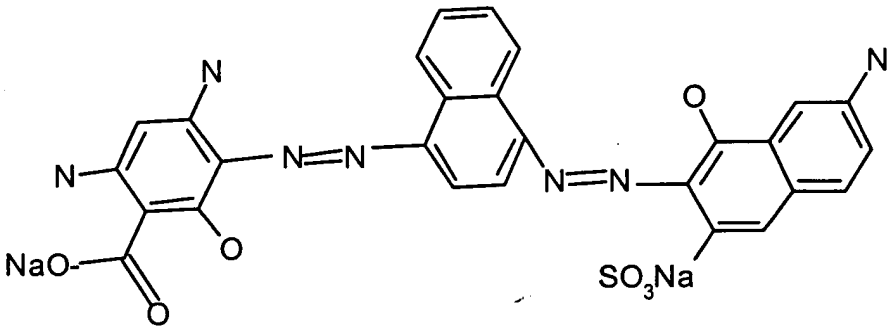
(DR1) Direct red 1; Sigma/Aldrich#S32,157-5

Fig.1H



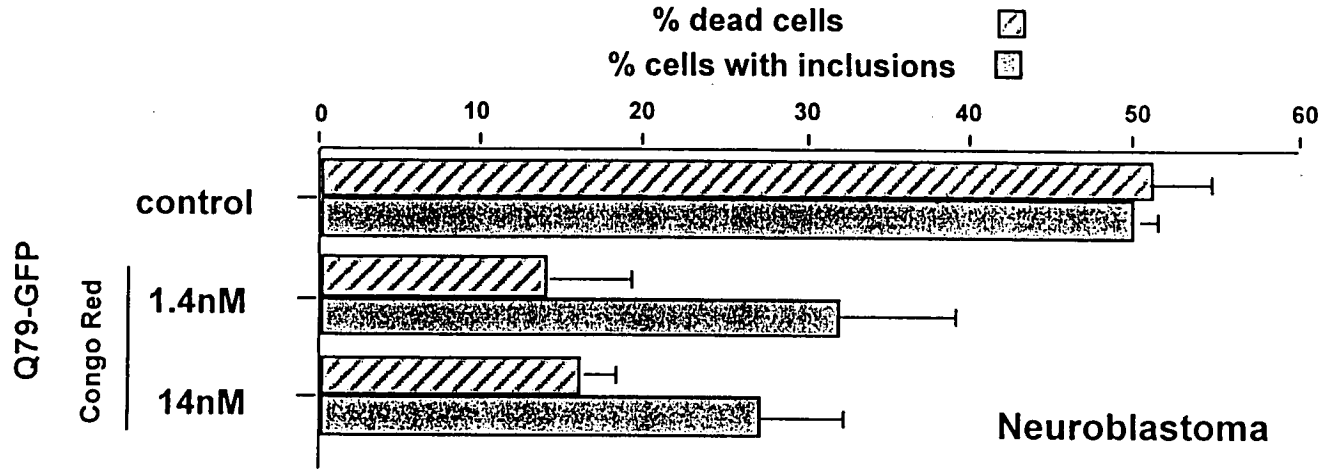
(DO1) Direct Orange 1; S33,0621

Fig.1I



(DB51) Direct Black 51; Sigma/Aldrich#S1,205-0

Fig.2A



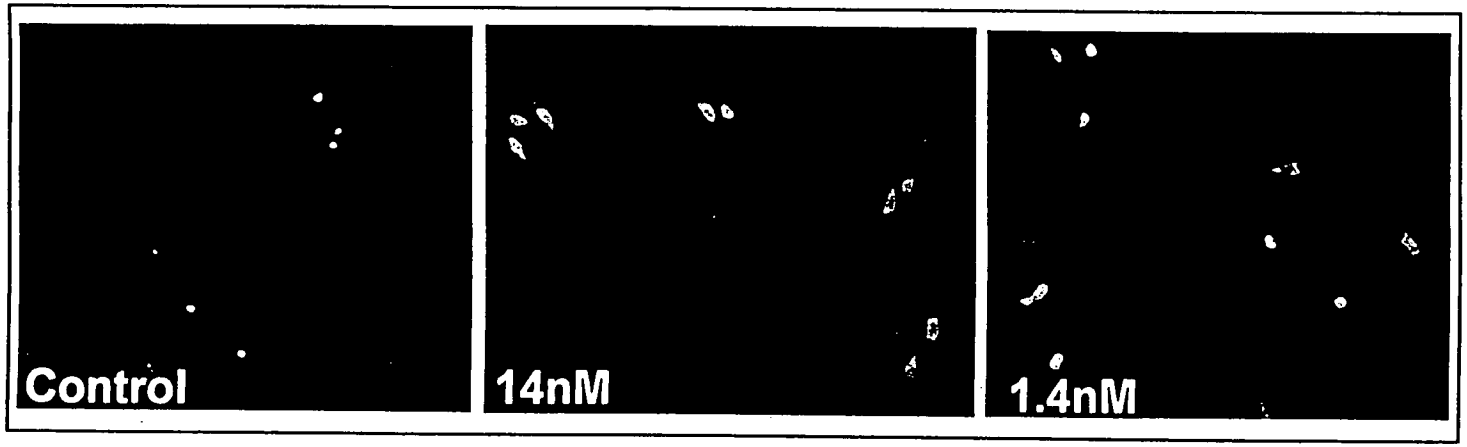


Fig.2B

Fig.2C

Fig.2D

Fig.3A

Fig.3B

Fig.3C

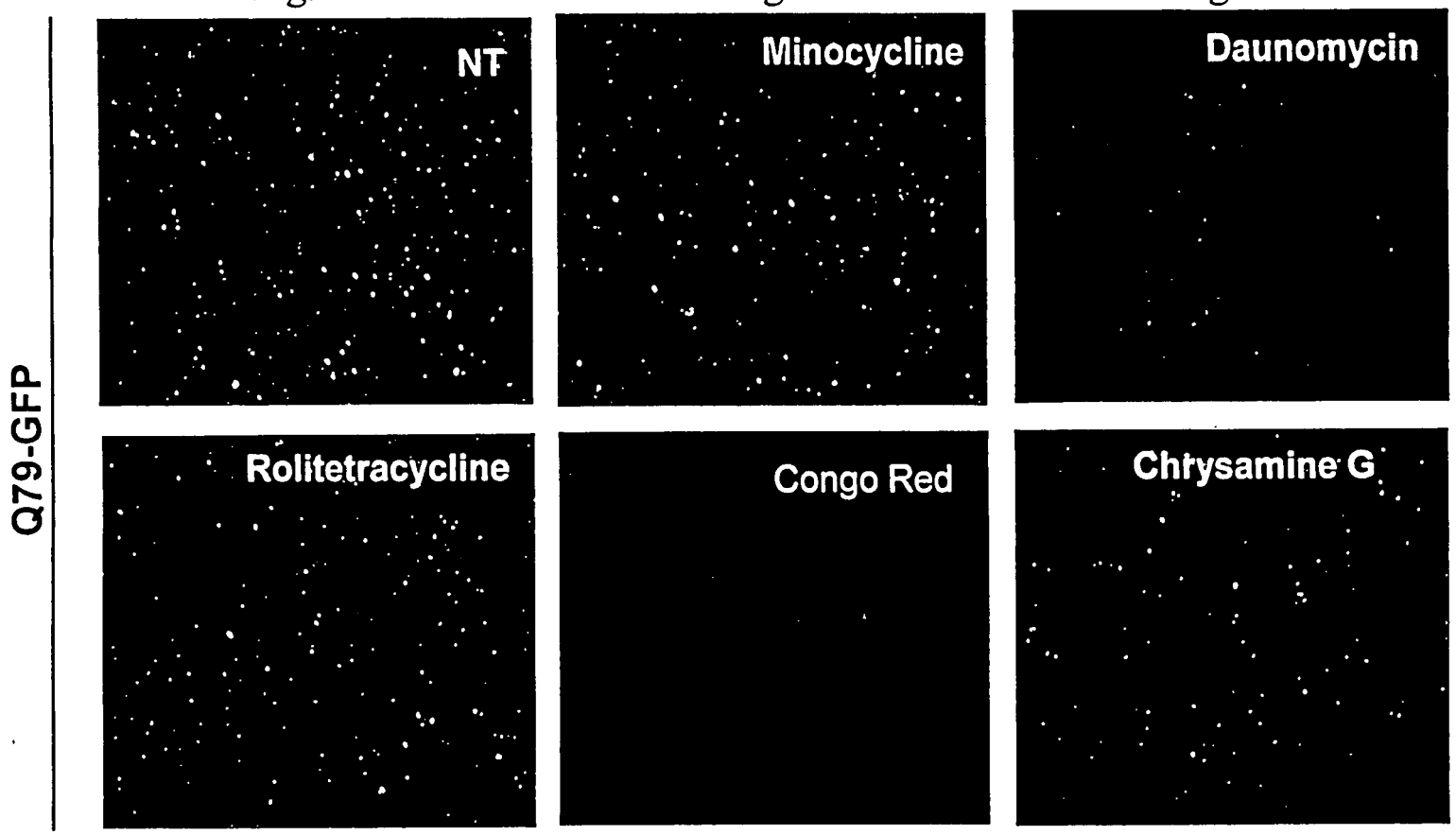
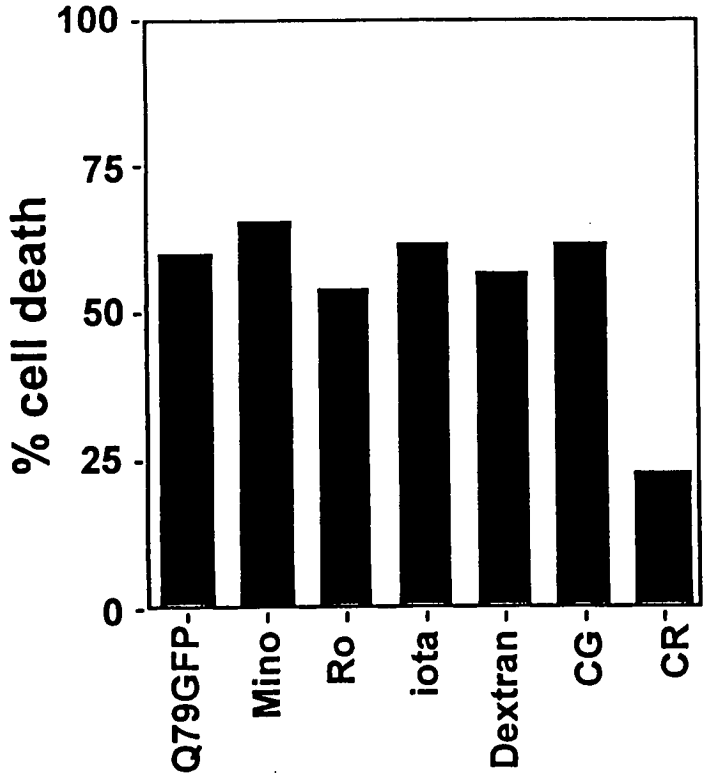


Fig.3D

Fig.3E

Fig.3F

Fig. 4A


100
75
50
25
0
% cell death
Q79GFP
Mino
Ro
iota
Dextran
CG
CR

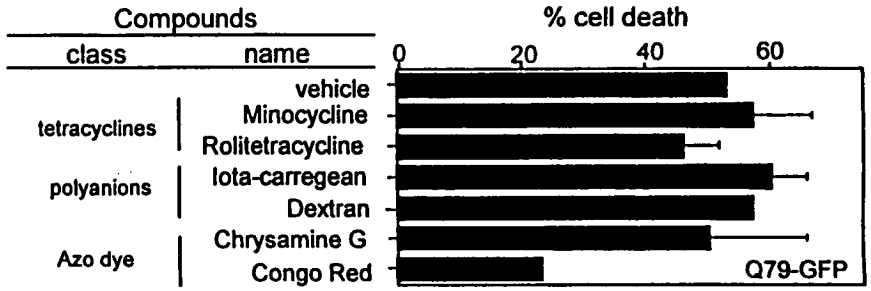

Compounds
class
name
% cell death
0
20
40
60
vehicle
tetracyclines
Minocycline
Rolitetracycline
polyanions
Iota-carregean
Dextran
Azo dye
Chrysamine G
Congo Red
Q79-GFP

Fig.4B

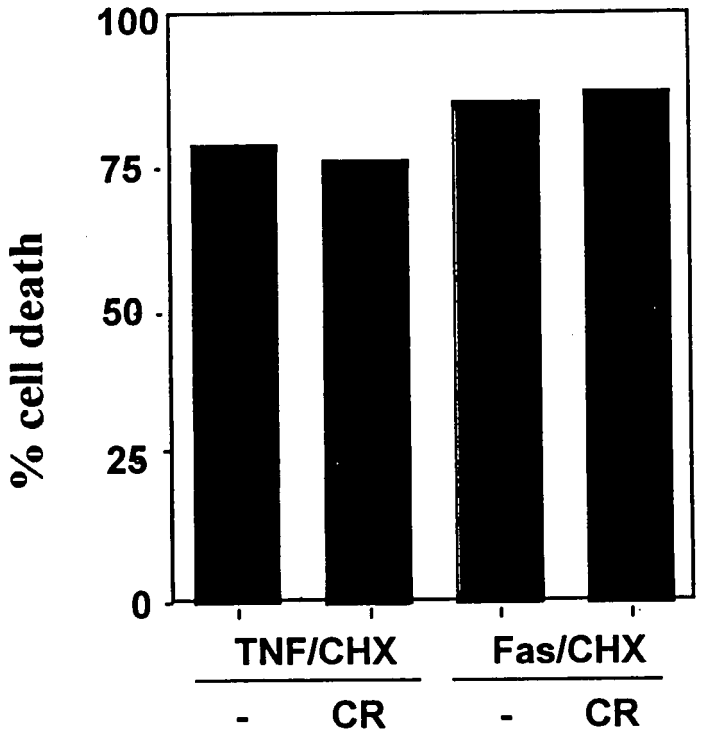
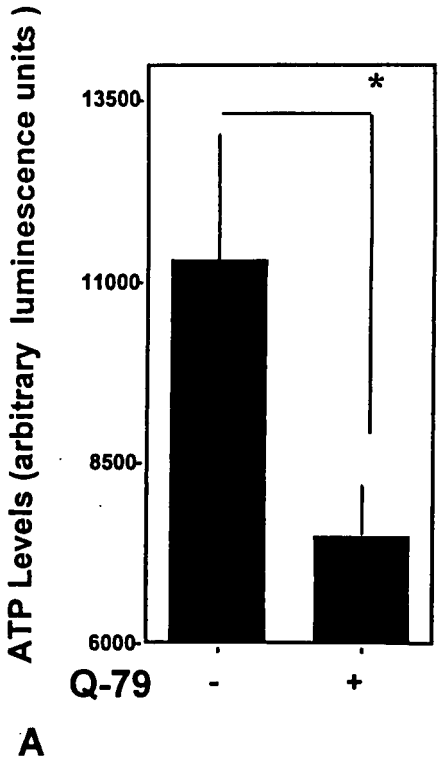


Fig.5A



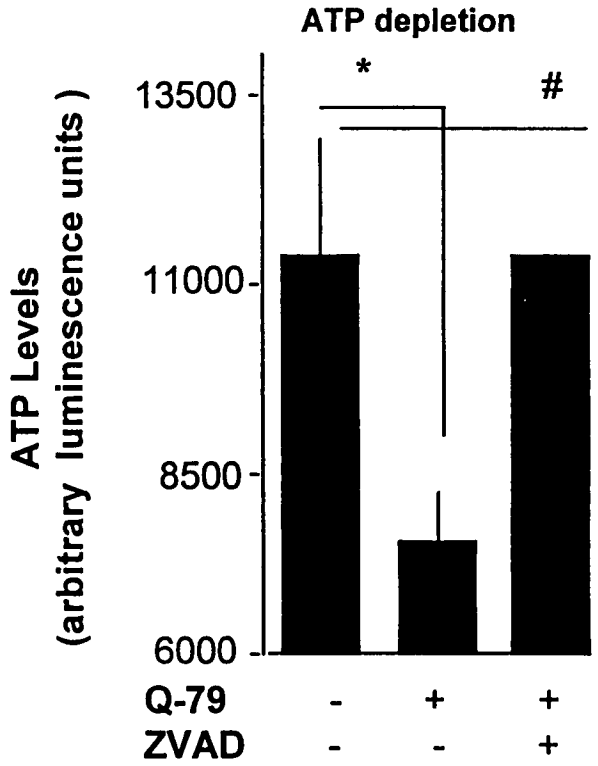


Fig. 5B

Fig. 5C

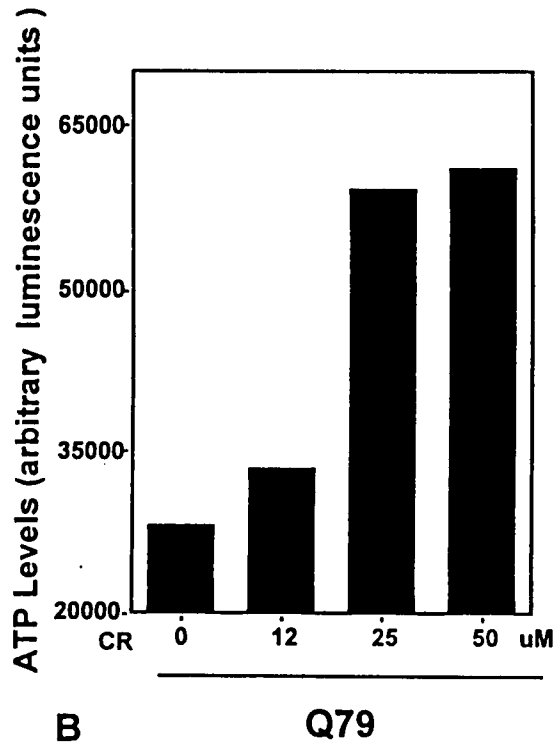

ATP Levels (arbitrary luminescence units)
65000
50000
35000
20000
CR
0
12
25
50
uM
B
Q79

Fig. 5D

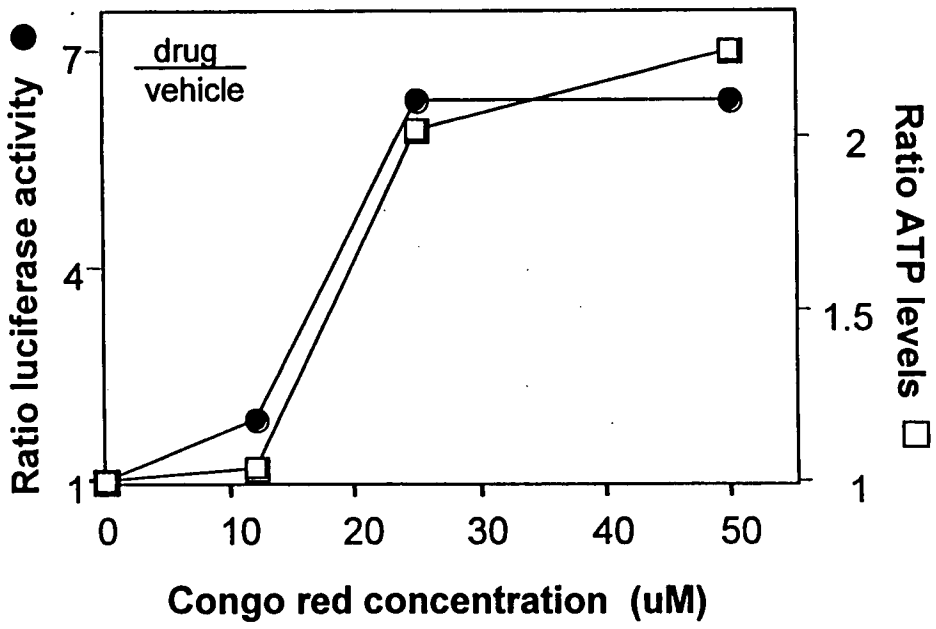

drug
vehicle
Ratio luciferase activity ●
Ratio ATP levels □
7
4
1
2
1.5
1
0
10
20
30
40
50
Congo red concentration (uM)

Fig. 5E

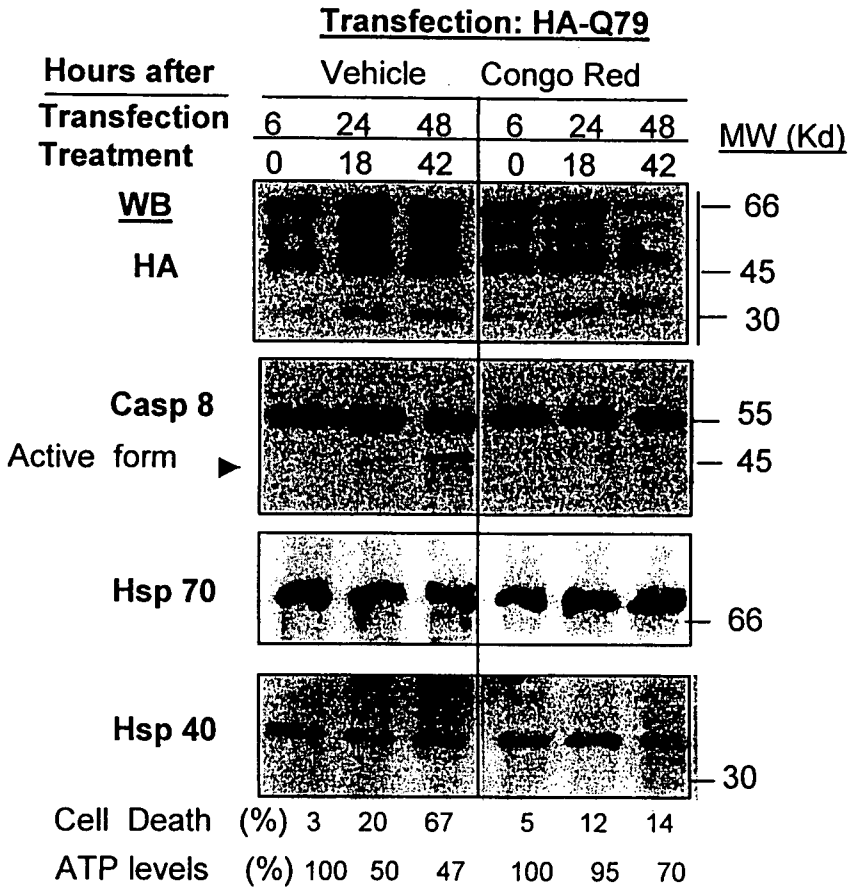

Transfection: HA-Q79
Hours after Transfection Treatment
Vehicle
Congo Red
6 24 48 6 24 48
0 18 42 0 18 42
MW (Kd)
WB
HA
66
45
30
Casp 8
Active form
55
45
Hsp 70
66
Hsp 40
30
Cell Death (%) 3 20 67 5 12 14
ATP levels (%) 100 50 47 100 95 70

Fig. 5F

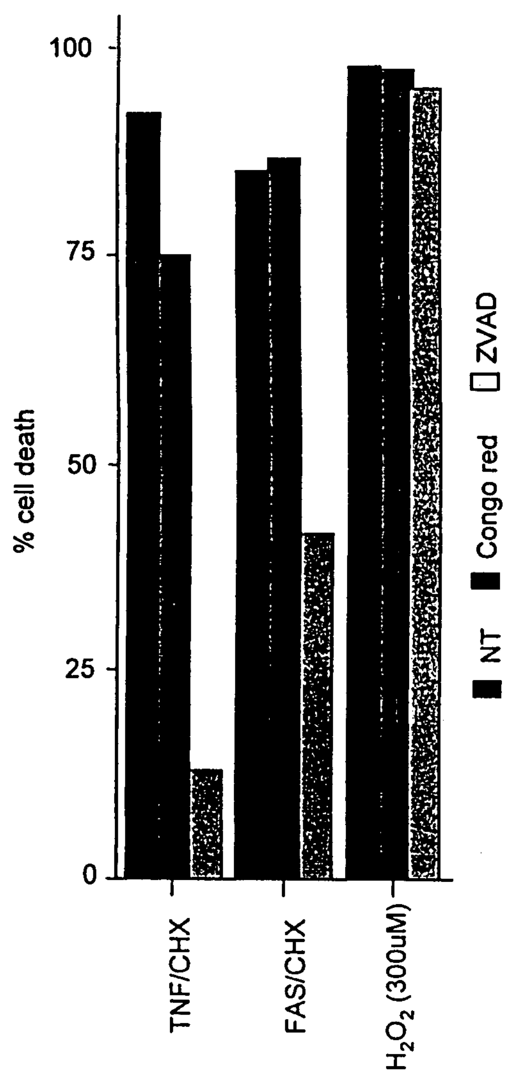


Fig. 5G

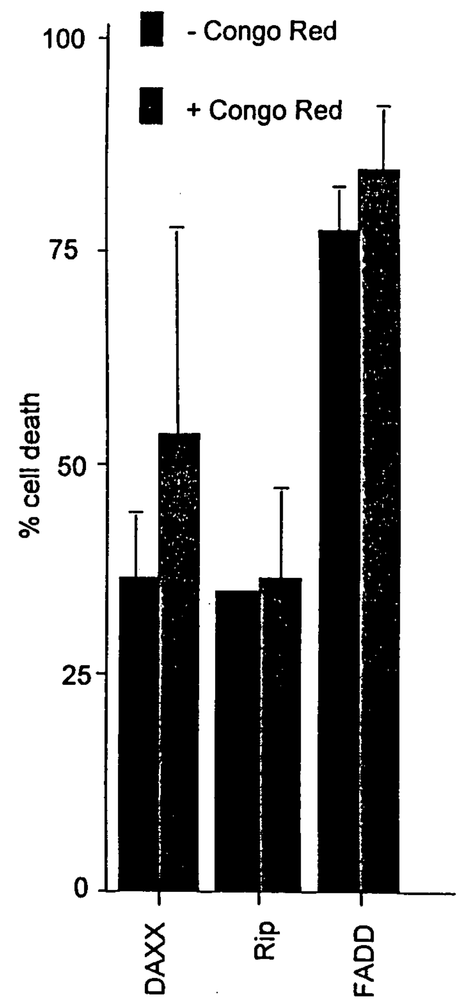
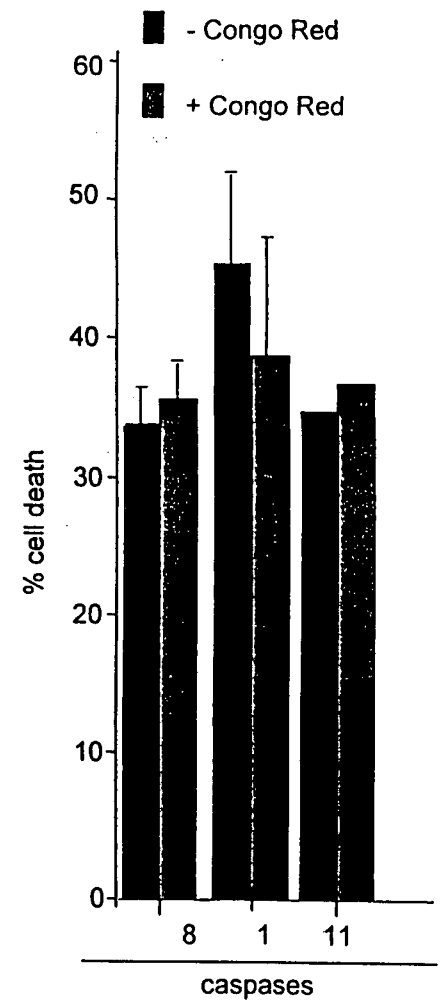


Fig. 5H



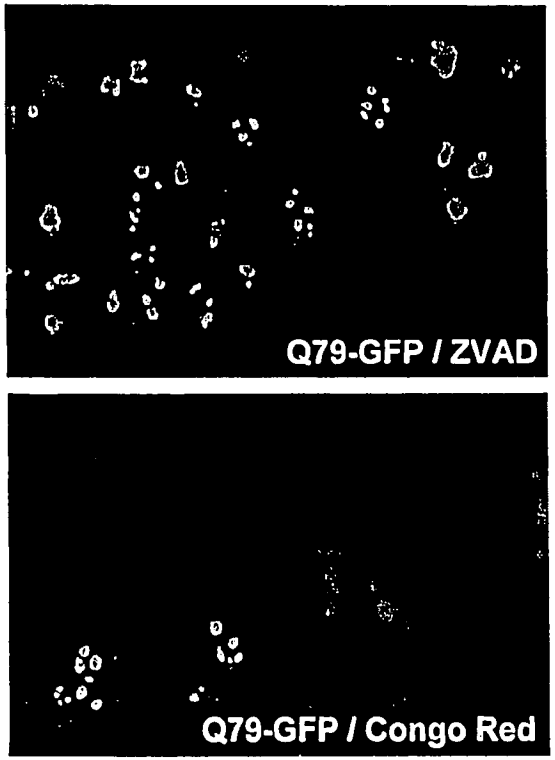


Fig. 5I

Fig. 6A

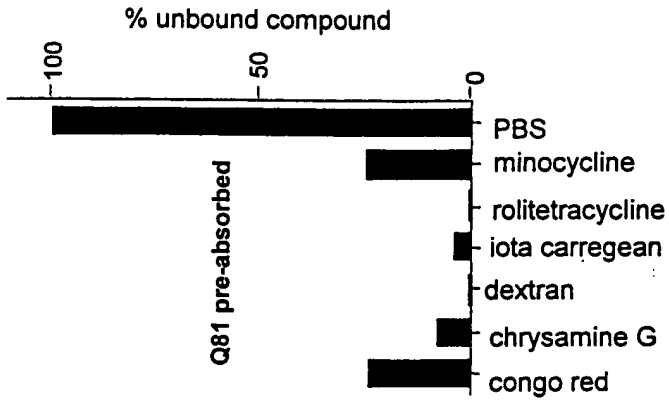

% unbound compound
100
50
0
Q81 pre-absorbed
PBS
minocycline
rolitetracycline
iota carregean
dextran
chrysamine G
congo red

Fig. 6B

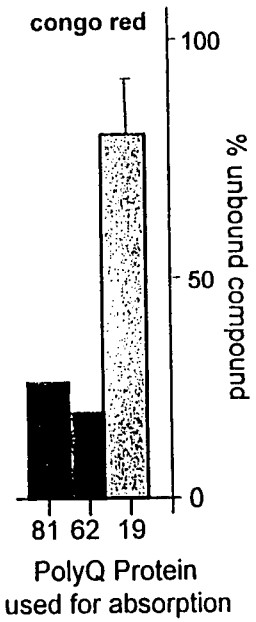
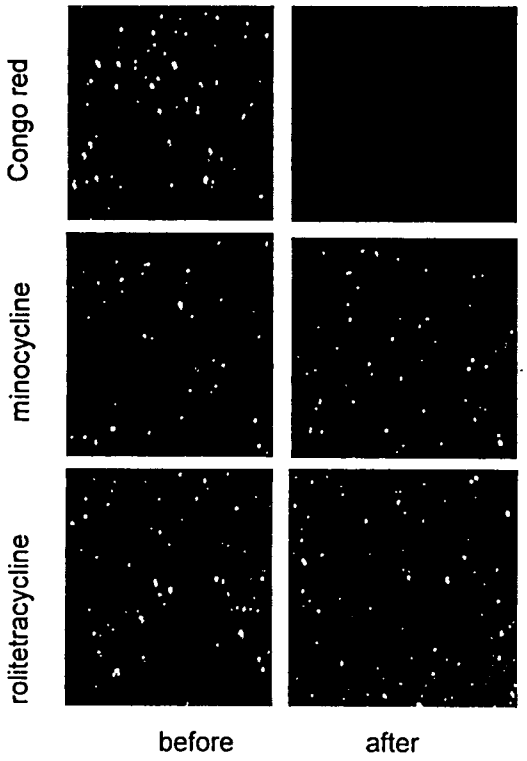


Fig. 6C



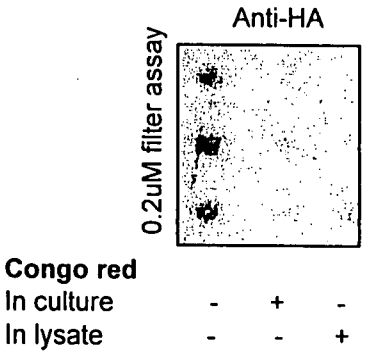


Fig. 6D

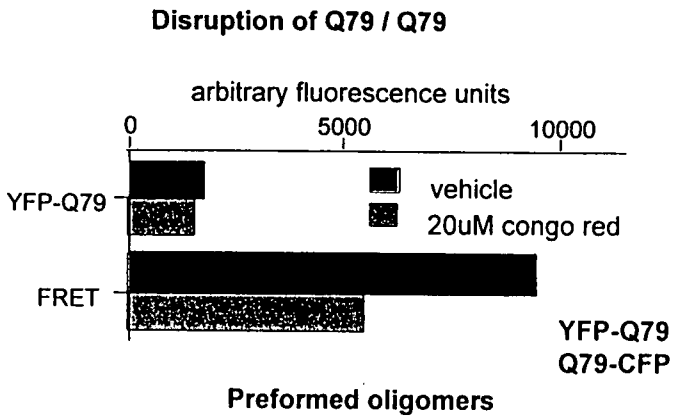
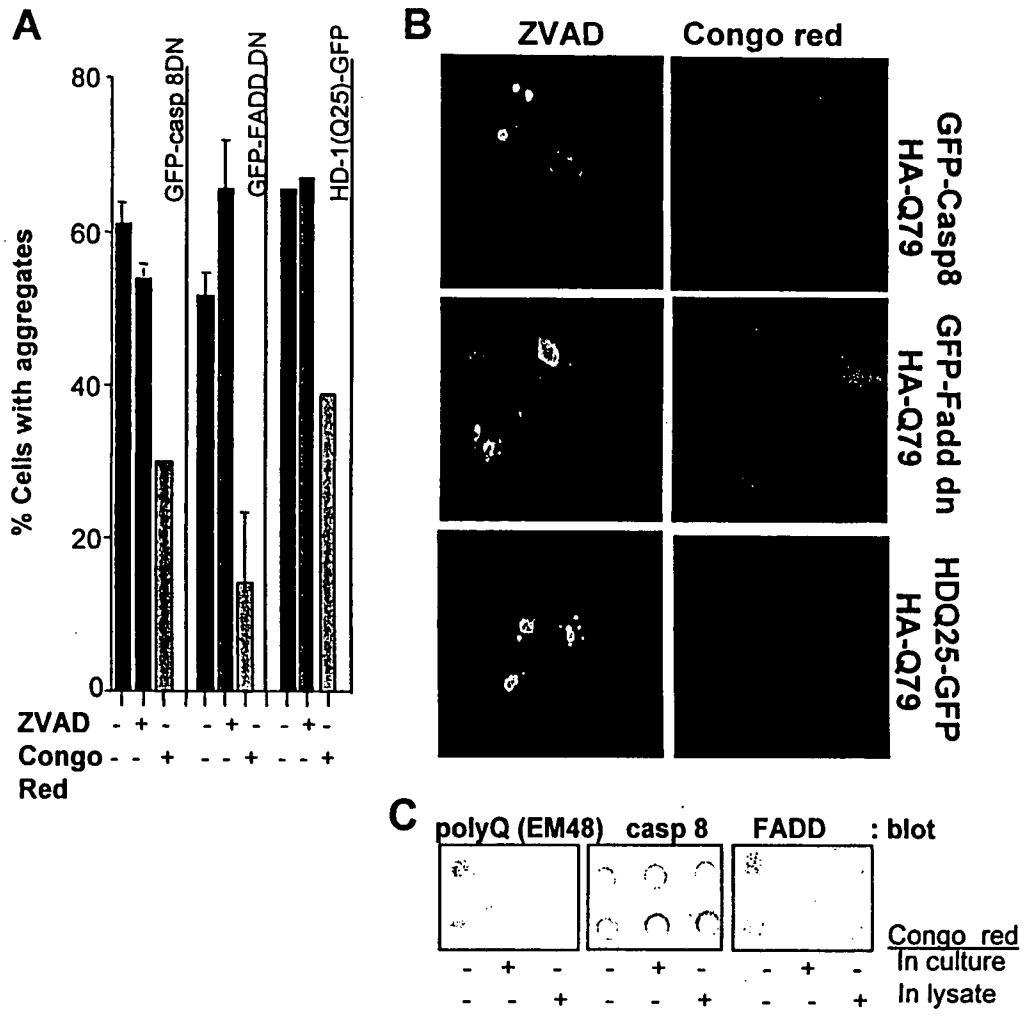
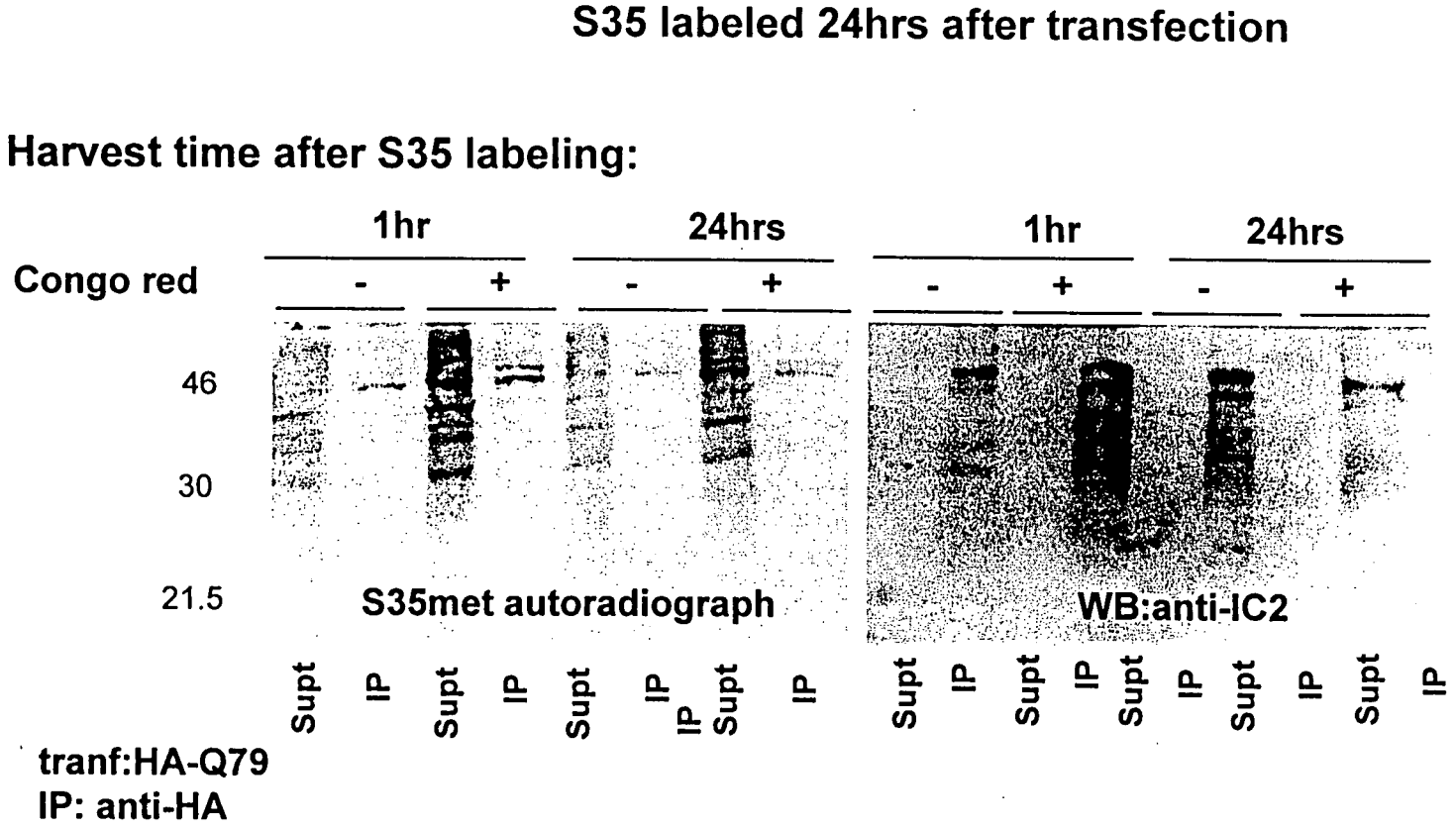


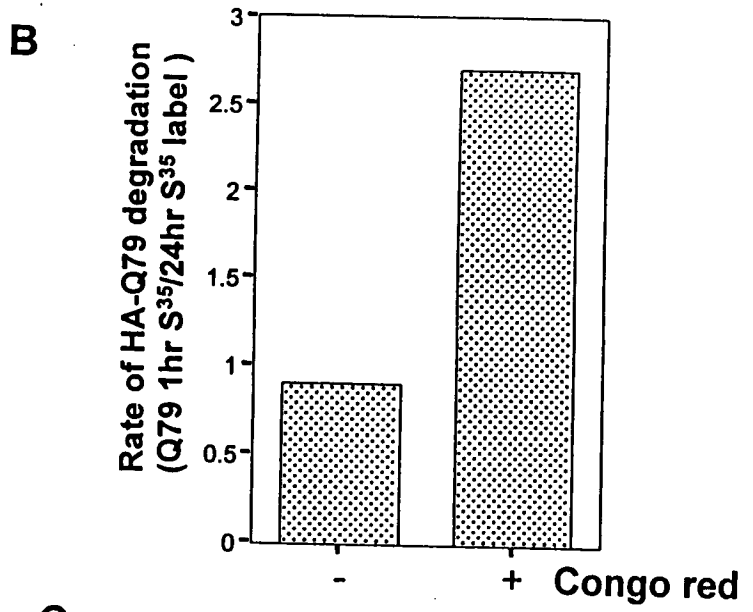
Fig. 6E

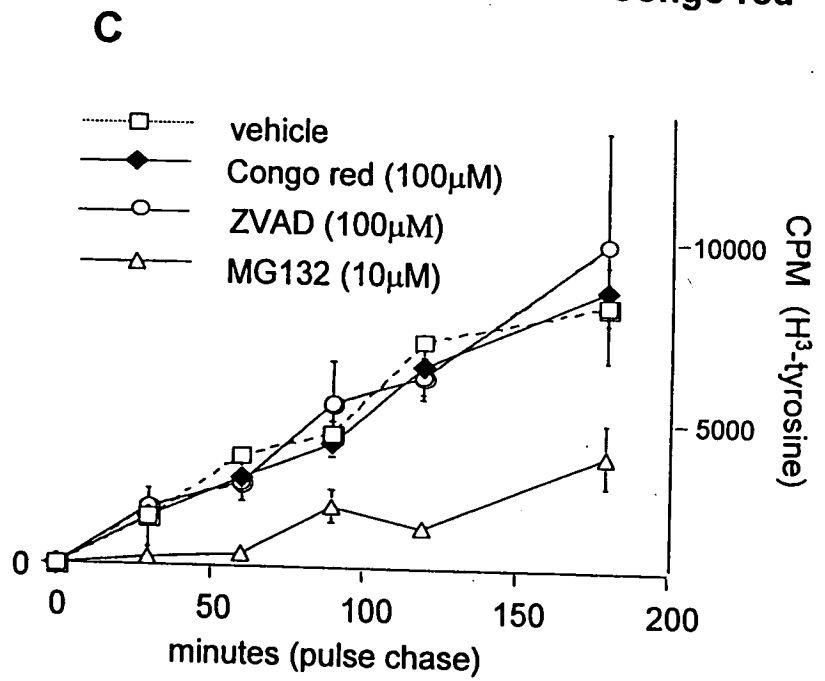

A
% Cells with aggregates
80
60
40
20
0
GFP-casp 8DN
GFP-FADD DN
HD-1(Q25)-GFP
ZVAD - + - - + - - + -
Congo - - + - - + - - +
Red
B
ZVAD
Congo red
GFP-Casp8 HA-Q79
GFP-Fadd dn HA-Q79
HDQ25-GFP HA-Q79
C
polyQ (EM48)
casp 8
FADD
: blot
Congo red
In culture
In lysate
- + - - + - - + -
- - + - - + - - +

Figs. 7A-C

Fig. 8A

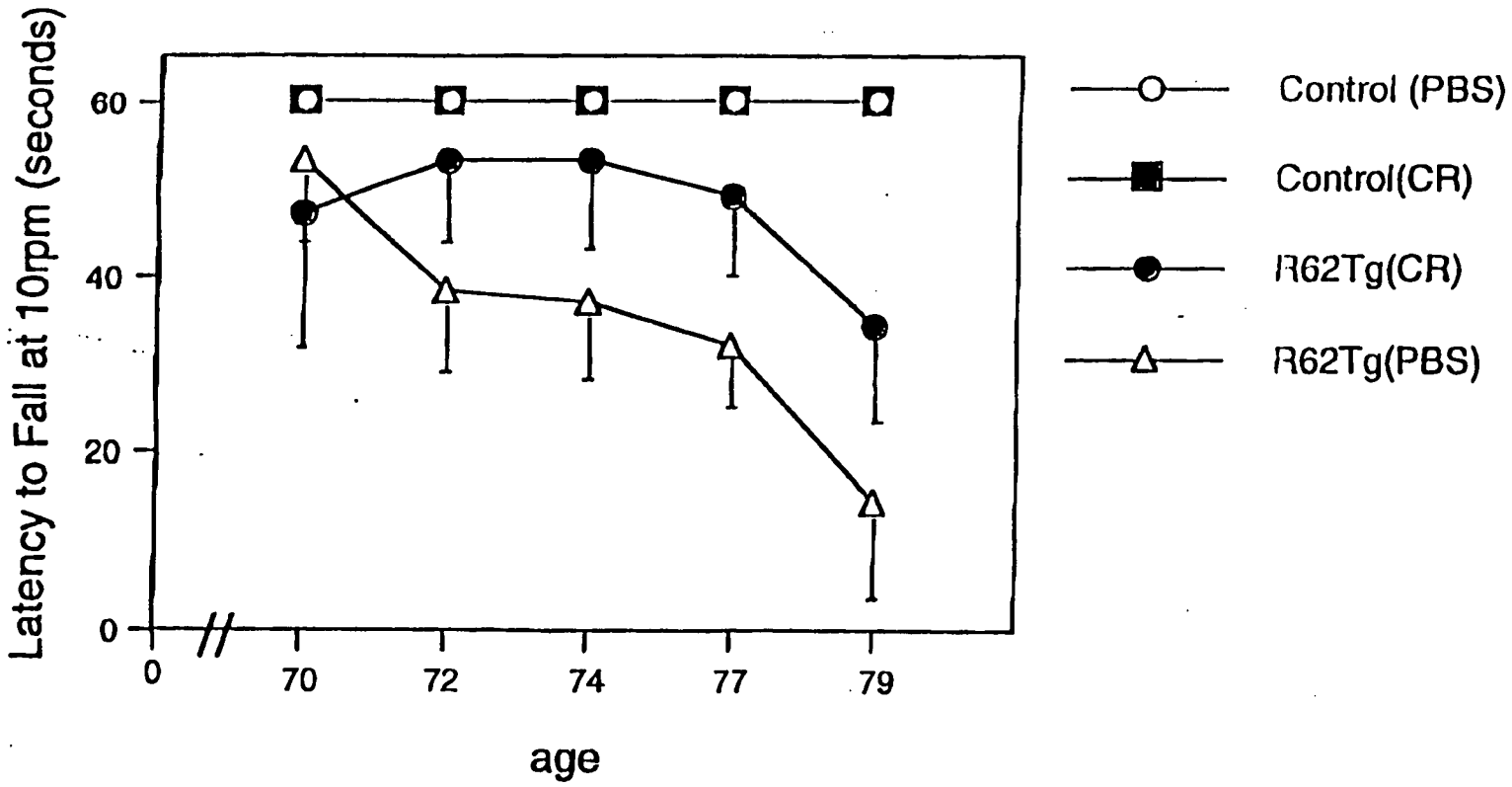

S35 labeled 24hrs after transfection
Harvest time after S35 labeling:
1hr
24hrs
1hr
24hrs
Congo red
- + - + - + - +
46
30
21.5
S35met autoradiograph
WB:anti-IC2
Supt IP Supt IP Supt IP IP Supt IP Supt IP Supt IP Supt IP Supt IP
tranf:HA-Q79
IP: anti-HA



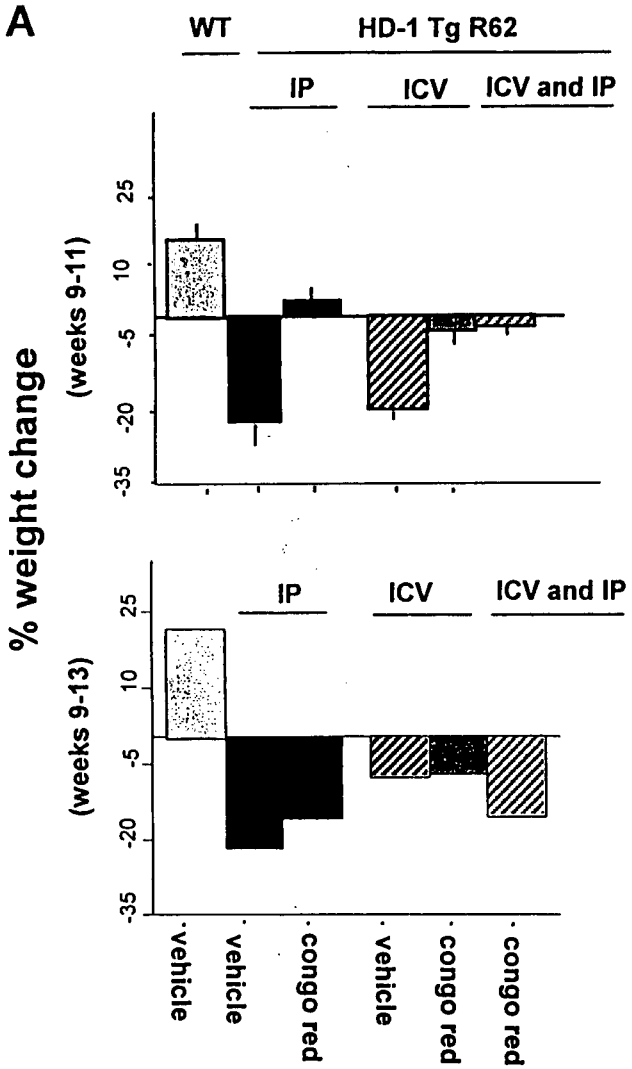


Figs. 8B & 8C

Fig. 9



A

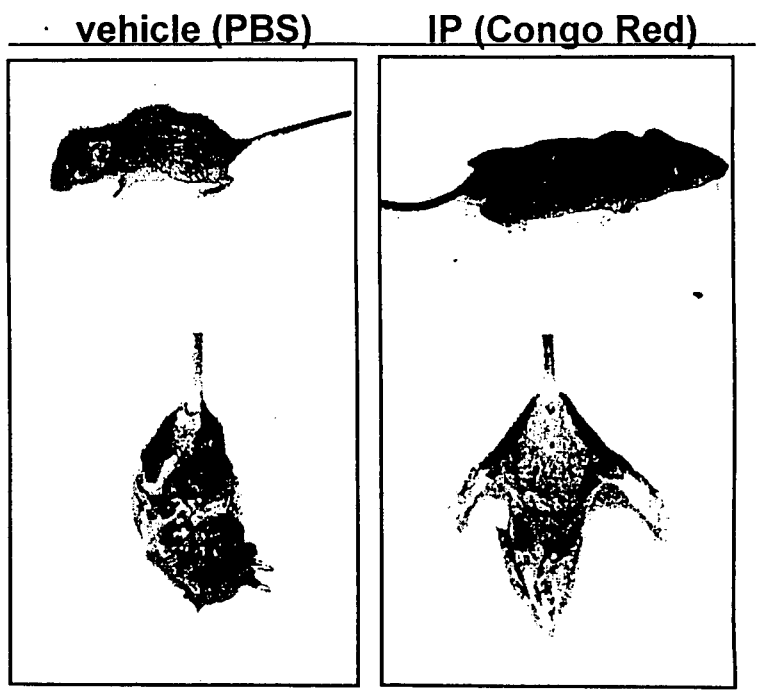


B

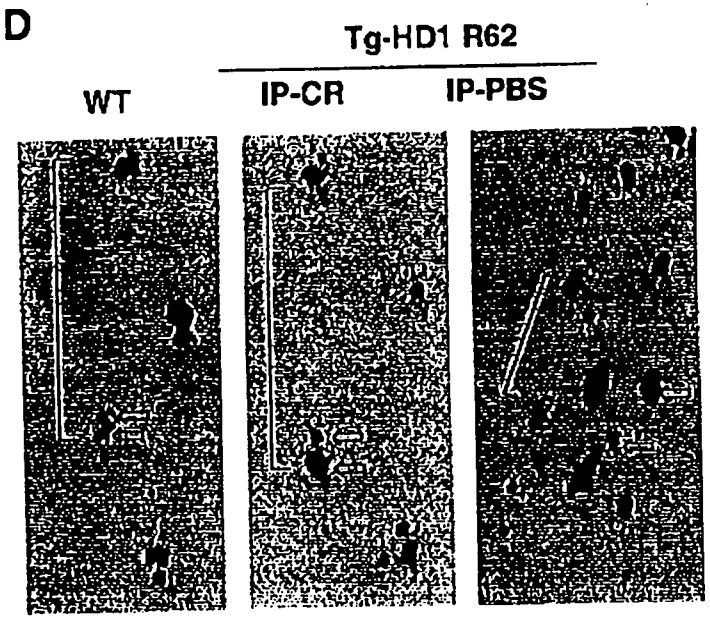
Fasting (6hrs) glucose levels

Wt	PBS-IP	152 mg/dl ± 27	
Wt	Congo Red-IP	216 mg/dl ± 60	p>0.05
HD1 Tg	PBS-IP	398mg/dl ± 98	p<0.01**
HD1 Tg	Congo Red-IP	178 mg/dl ± 46	p>0.05, *p<0.05

C

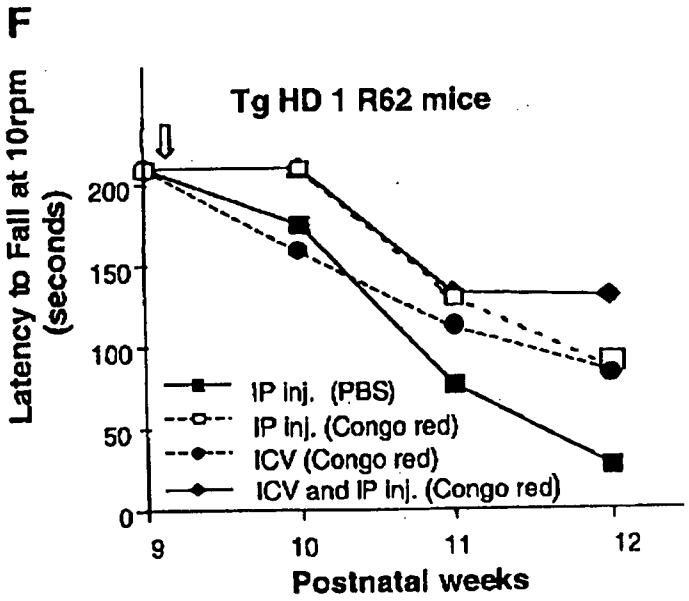


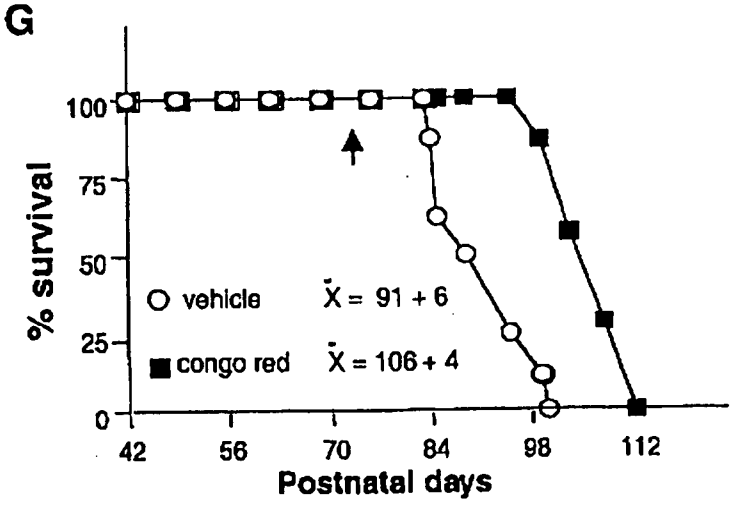
Figs. 10A - 10C



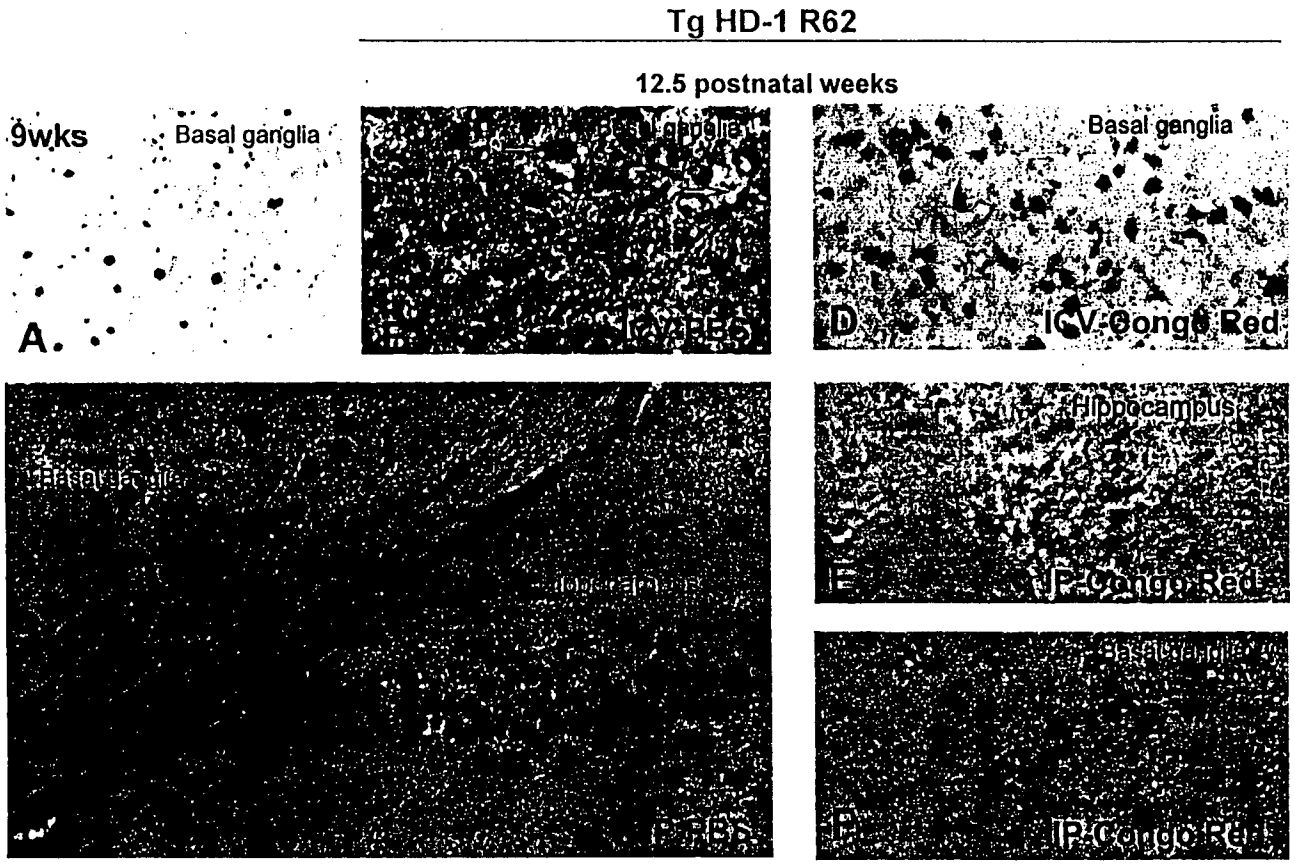
E

	Tg-HD1 R62-IP inj		
	vehicle	Congo red	
Stride:	-46%	-17 %	p<0.01



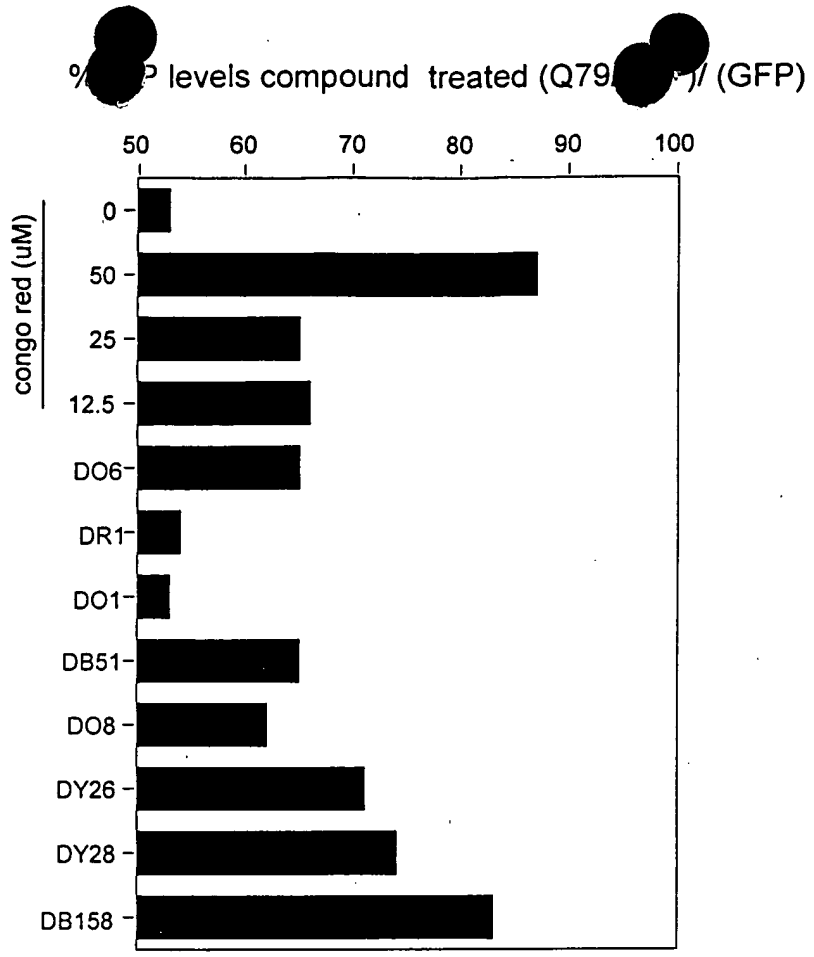


Figs. 10D - 10G

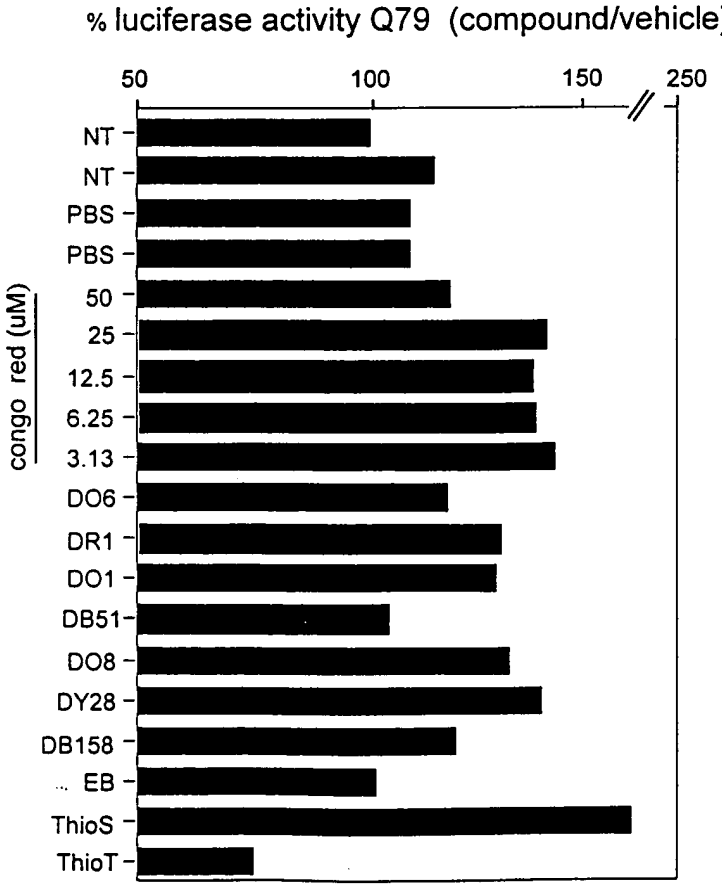


Figs. 11A-11F

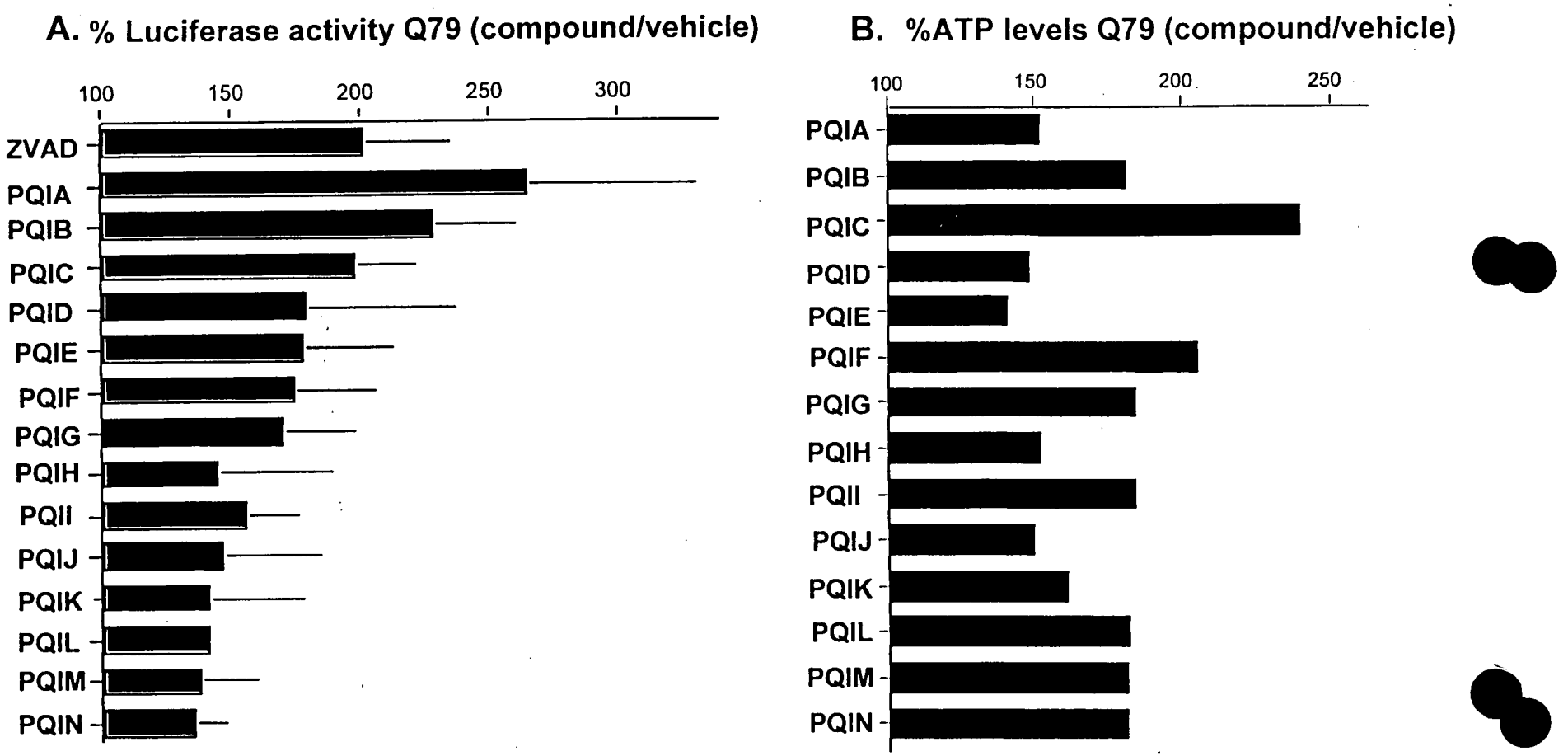
A



B

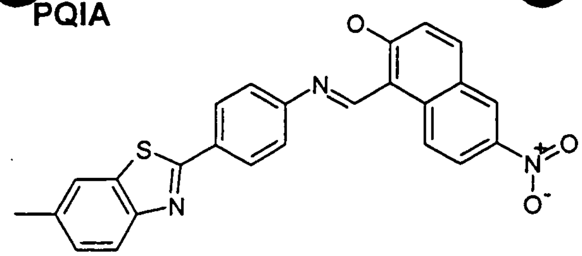


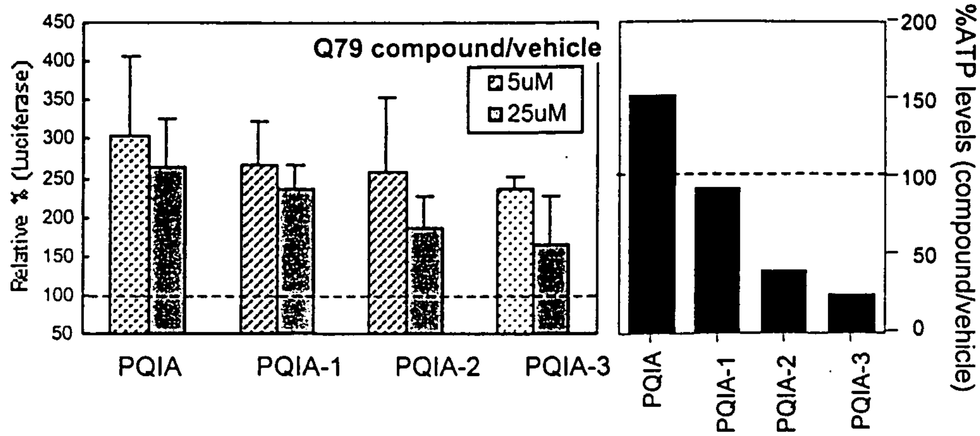
Figs. 12A & 12B

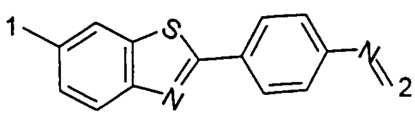


Figs. 13A & 13B

PQIA



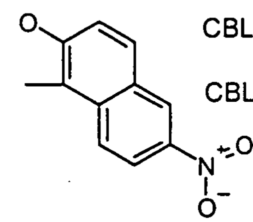


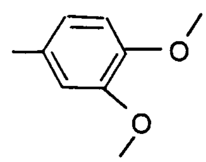


PQIA

PQIA : 1=CH_3 2= CBL#250313

PQIA-1 : 1=H 2= CBL# 250329



PQIA-2 : 1=CH_3, 2=  CBL#191895

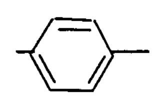
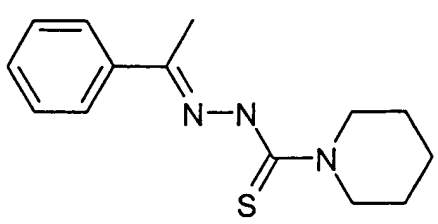
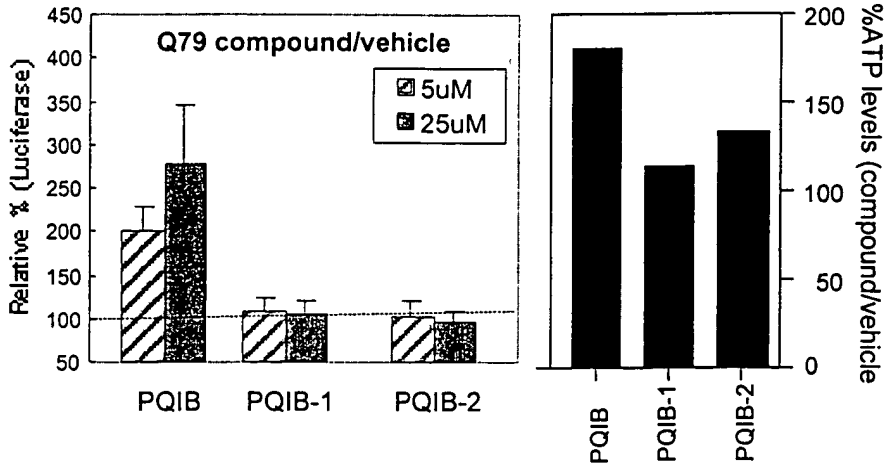
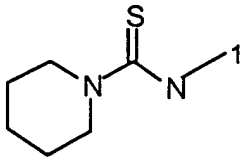
PQIA-3 : 1=CH_3 2=  CBL#191886

Fig. 14A

PQIB

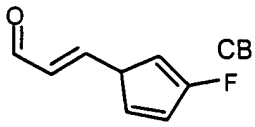






PQIB

PQIB : CBL#285042

PQIB-1 : 1=  CBL#150931

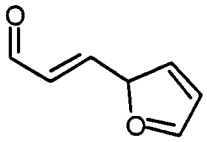
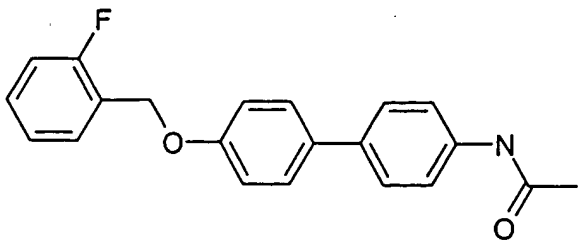
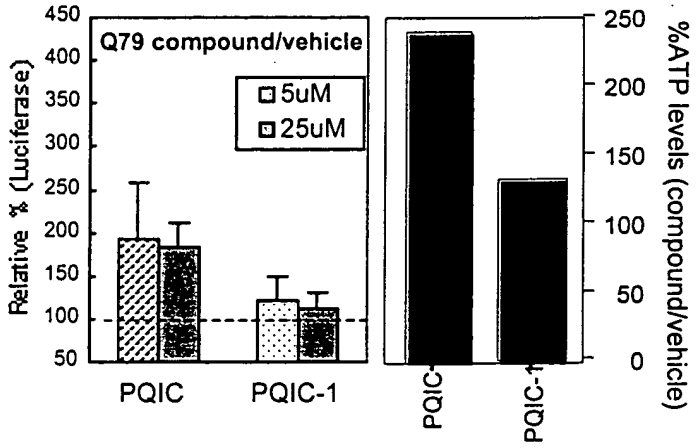
PQIB-2 : 1=  CBL#150873

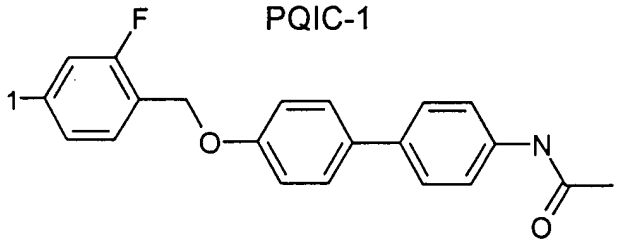
Fig. 14B

PQIC





PQIC-1



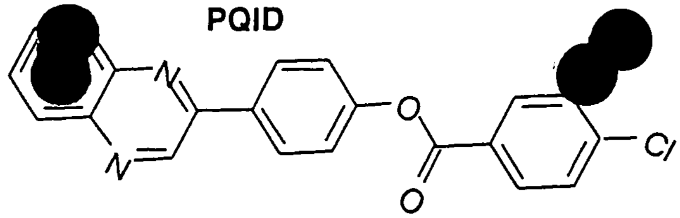
PQIC

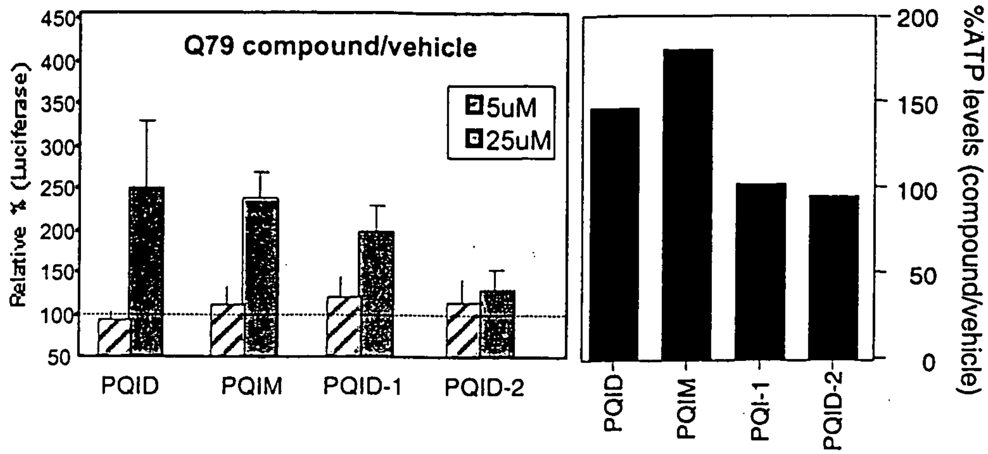
PQIC : 1 = H CBL#243678

PQIC-1 : 1= NO_2- CBL#243676

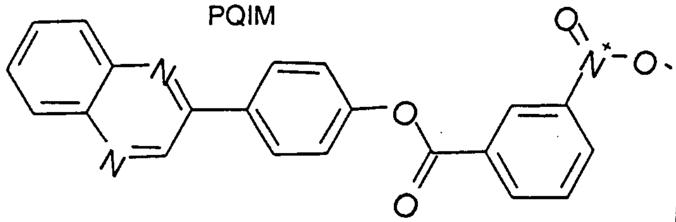
Fig. 14C

PQID

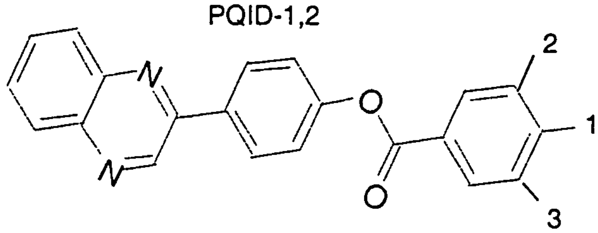




PQIM



PQID-1,2

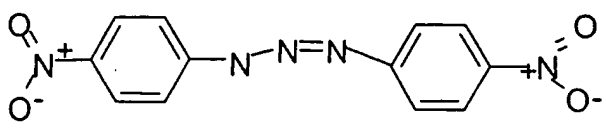


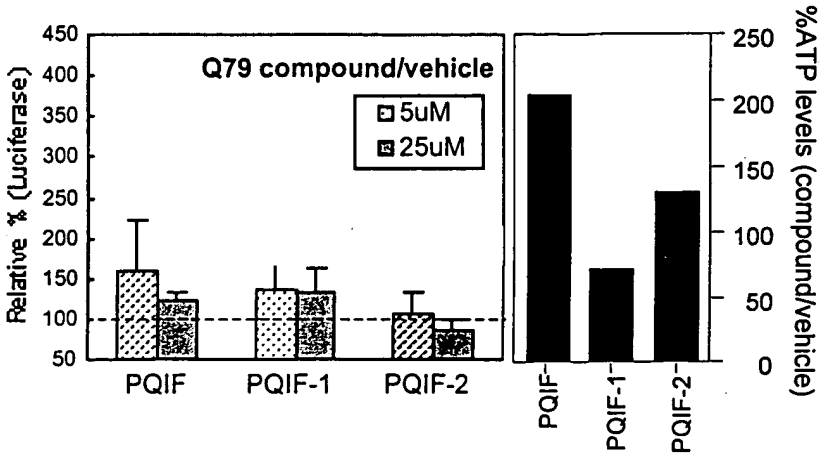
<u>PQID</u>

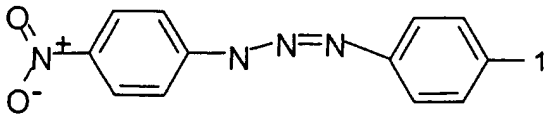
PQID :	1 = Cl, 2 =H, 3 =H	CBL#163012
PQIM :	1=H, 2 = NO_2, 3 =H	CBL#162753
PQID-1 :	1=Br, 2=H, 3= NO_2	CBL#162728
PQID-2 :	3= Br 2=H 3=H	CBL#162759

Fig. 14D

PQIF







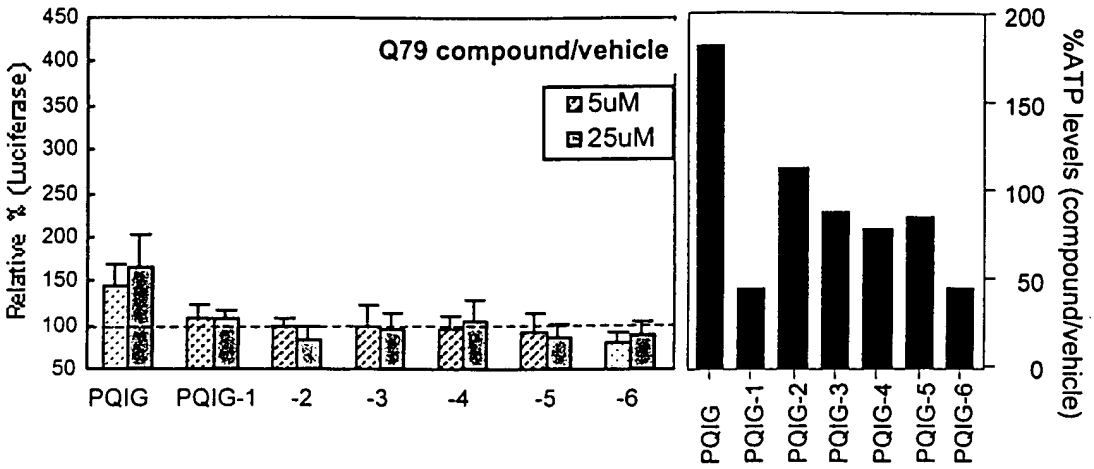
PQIF-1,2

PQIF

PQIF	:	1= NO_2	CBL#100707
PQIF-1	:	1=Br	CBL#122267
PQIF-2	:	1=O_2	CBL#136395

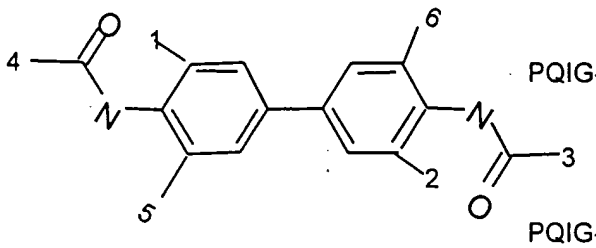
Fig. 14E

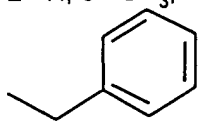
PQIG

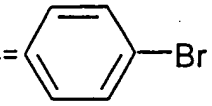


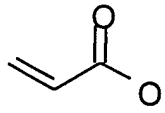
PQIG

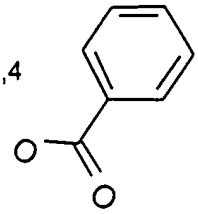
PQIG : 1= H, 2= H, 3= CH_3, 4= CH_3, 5= H CBL#104413

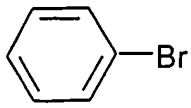


PQIG-1 : 3, 4=  1,2,5,6=H CBL#104234

PQIG-2 : 1,2=CH_3 , 3,4=  5,6= H CBL#116574

PQIG-3 : 3,4=  1,2= H 5,6= CH_3 CBL# 105109

PQIG-4 : 3,4  1,2,5,6=H CBL#105560

PQIG-5: 3,4=  1,2,5,6=H CBL#116580

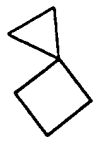
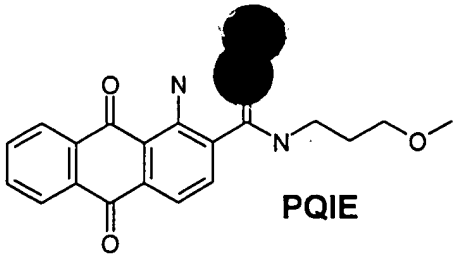
PQIG-6: 3,4=  1, 2, 5, 6= H CBL#153040

Fig. 14F

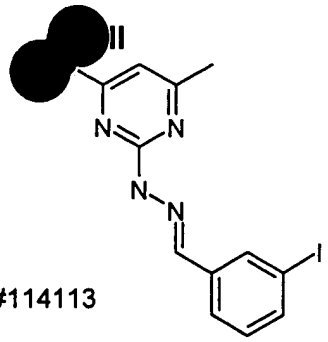
G



PQIE

CBL#249429

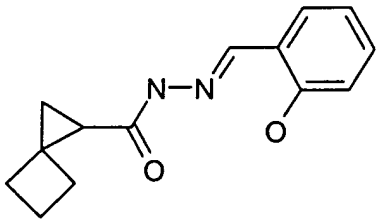
J



CBL#114113

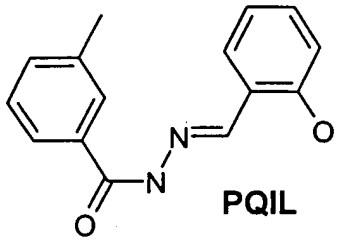
H

PQIK



CBL#269475

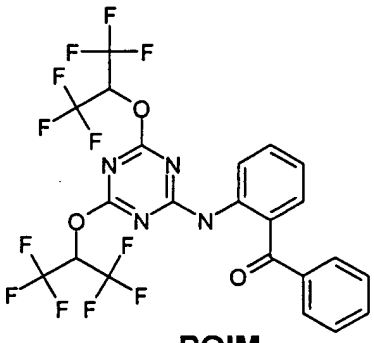
K



PQIL

CBL#261511

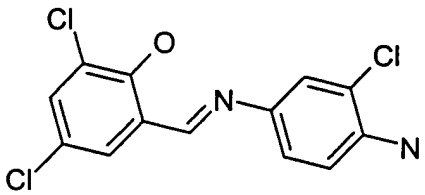
I



PQIM

CBL#162753

L



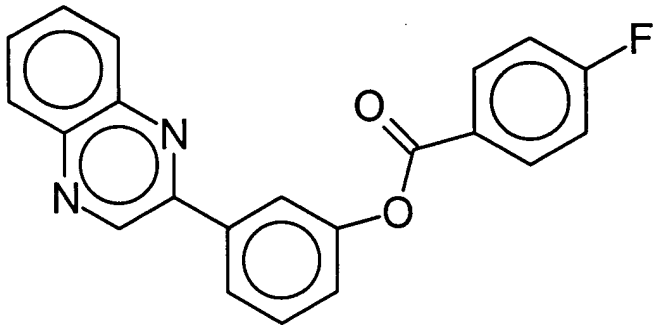
PQIN-1

CBL#267402

Figs. 14G - 14L

1: CNC-43921

Fig. 15A



#2: CNC-43267

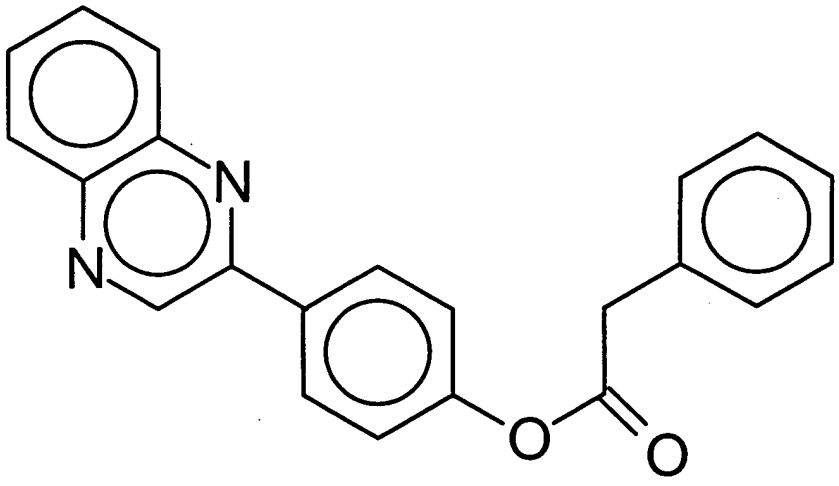


Fig. 15B

CNC - 49867

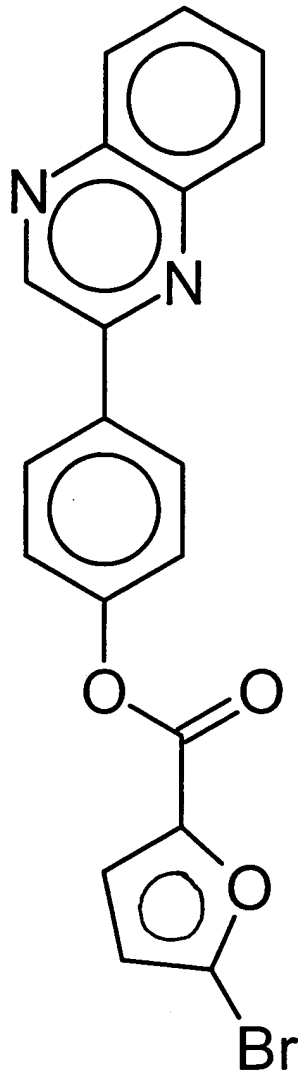


Fig. 15C

4 CNC-49867

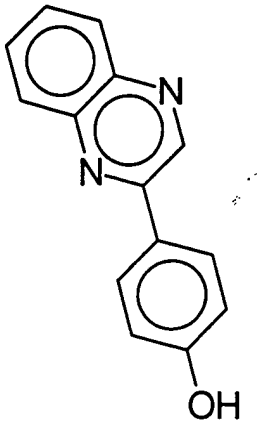


Fig. 15D

#5: CNC - 54580

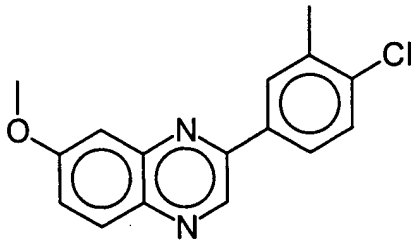


Fig. 15E

#6: CNC - 42175

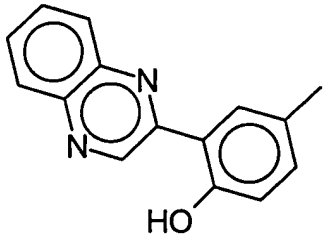


Fig. 15F

#7: CNC-42379

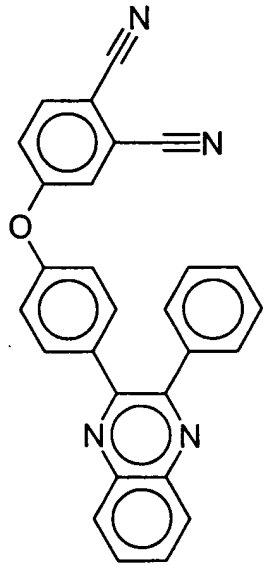


Fig. 15G

#8: CNC-46308

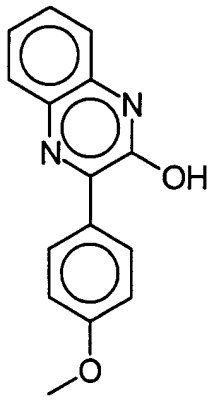


Fig. 15H

#9: CNC-46793

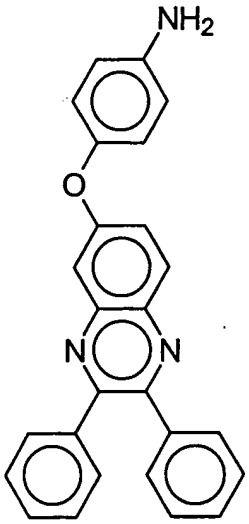


Fig. 15I

10: CNC-49373

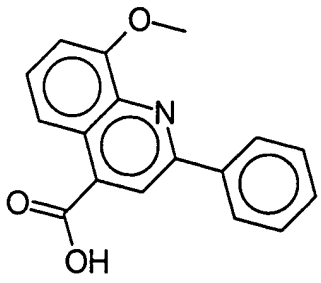
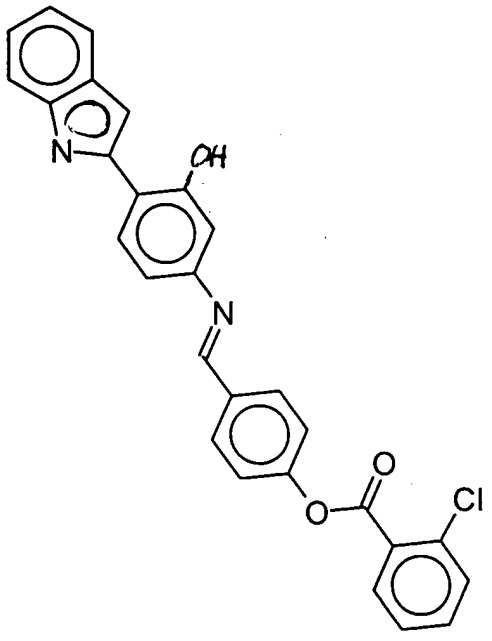


Fig. 15J

Fig. 15K

1: CNC-57277



#1: CNC-556240

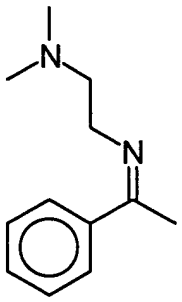


Fig. 15L

2 CNC-526900

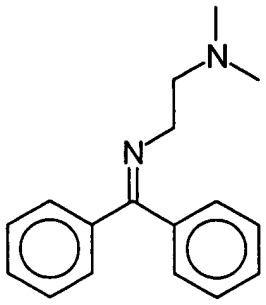
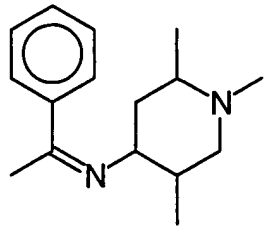


Fig. 15M

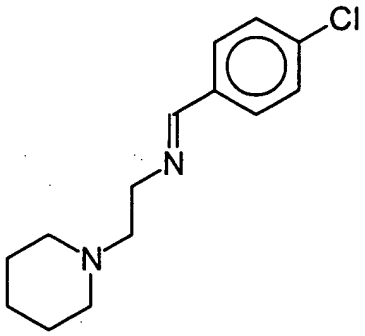
3: CNC-431893

Fig. 15N



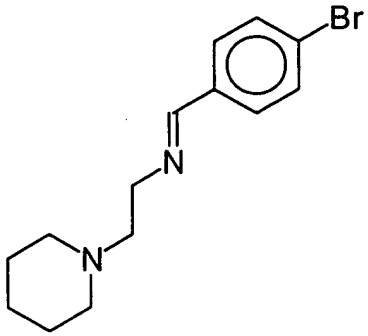
#4 : CNC-523618

Fig. 15O



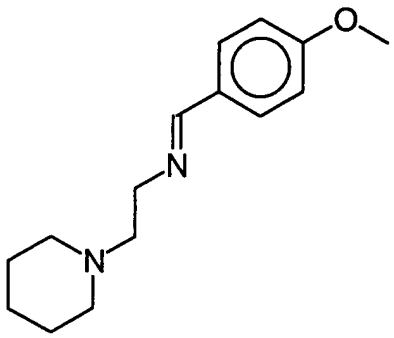
#5 : CNC-555148

Fig. 15P



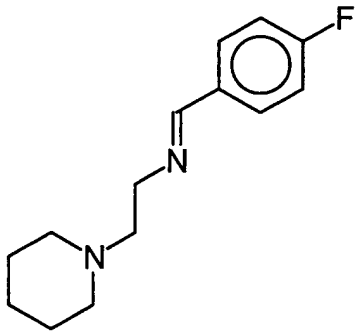
6: CNC-521484

Fig. 15Q



#7: CNC-543738

Fig. 15R



#8: CNC-529717

Fig. 15S

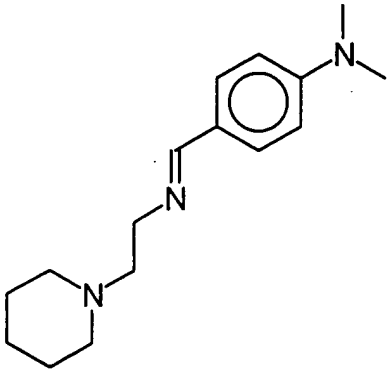
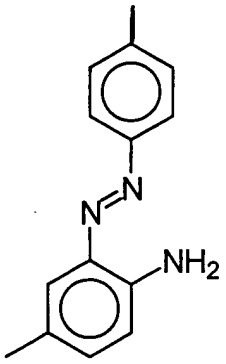


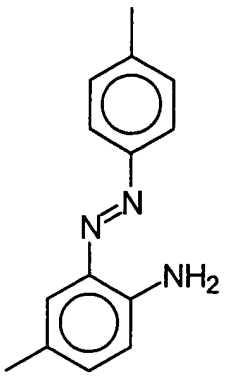
Fig. 16A

#1: CNC-289284



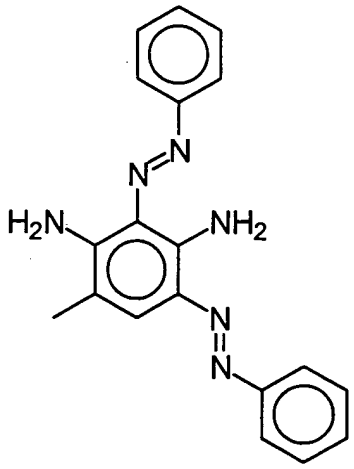
#2 : CNC-106942

Fig. 16B



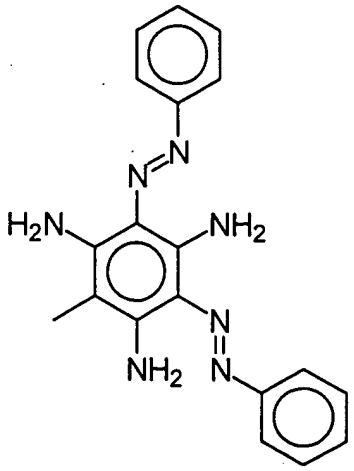
#3: CNC-287671

Fig. 16C



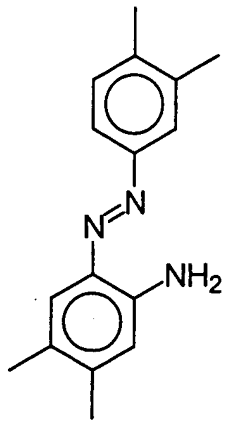
#4 : CNC-287227

Fig. 16D



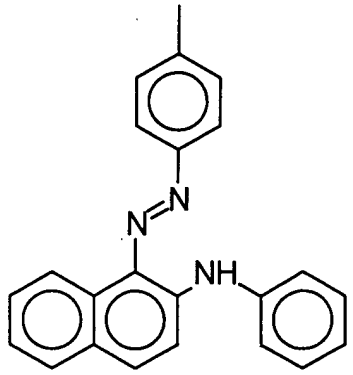
#5 : CNC-300273 **and CNC - 1268328**

Fig. 16E



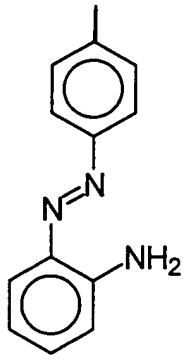
#7 : CNC-1308309

Fig. 16F



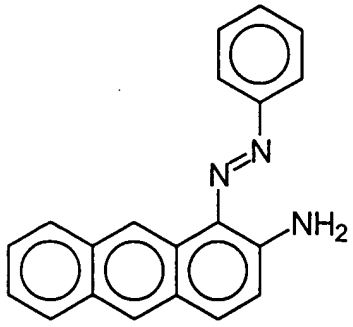
#8 : CNC-1069226

Fig. 16G



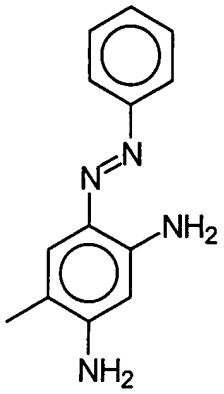
#9 : CNC-290524

Fig. 16H



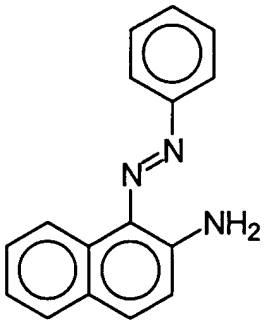
10 : CNC-609843

Fig. 16I



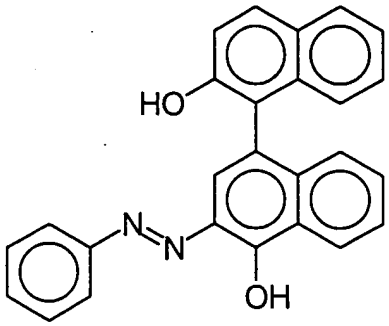
#11 : CNC-1059876

Fig. 16J



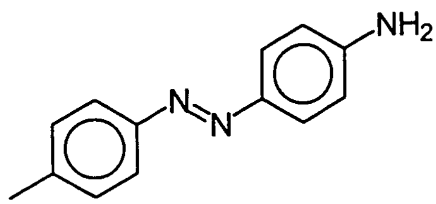
#12: CNC-300196

Fig. 16K



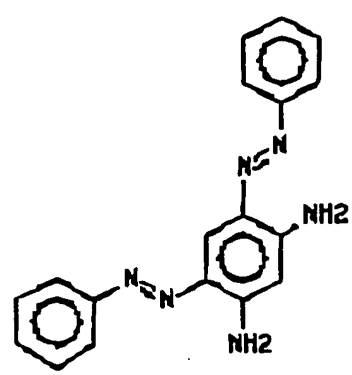
#13: CNC-287437

Fig. 16L

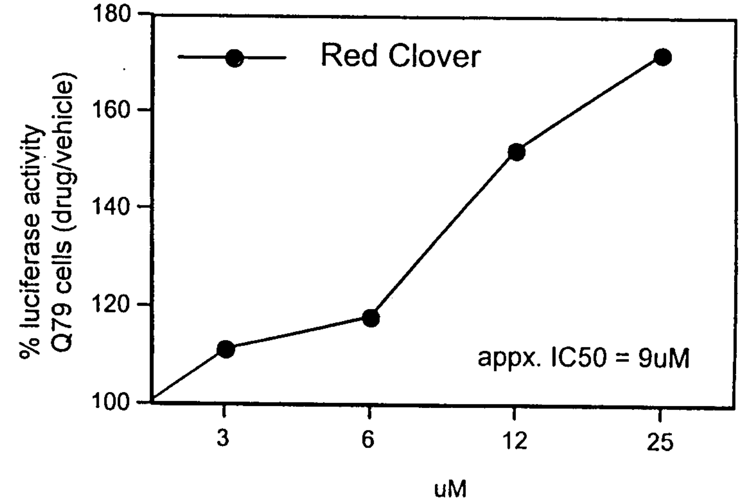


14 CNC-301181
15 CNC-628178
#16: CNC-1292419

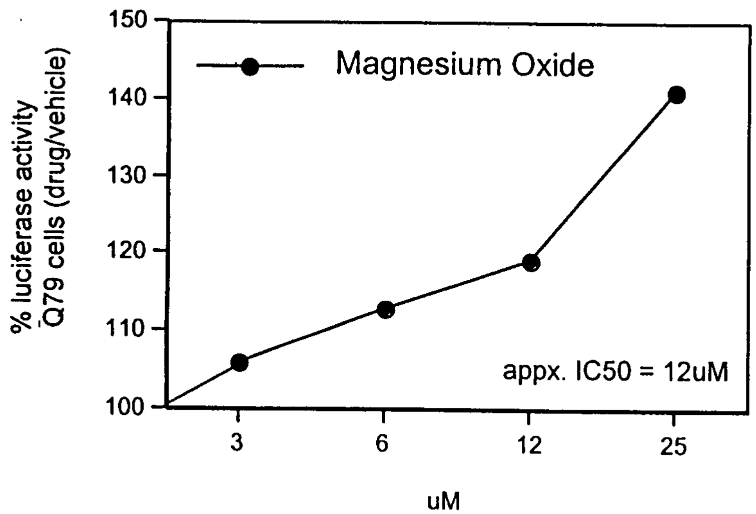
Fig. 16M



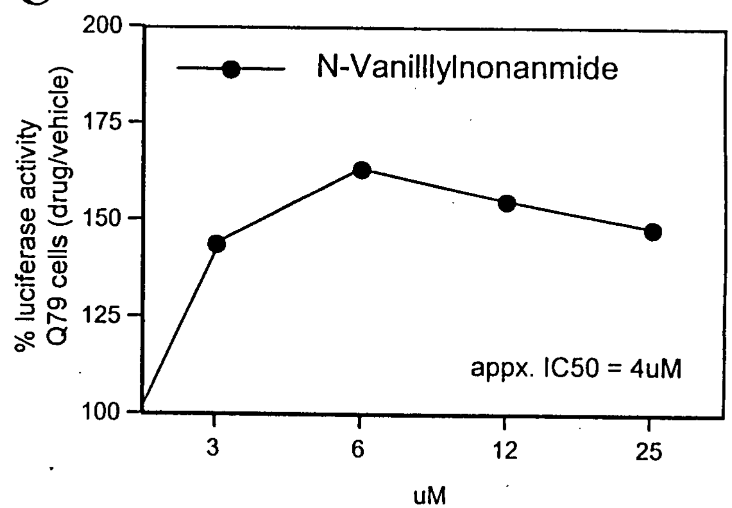
A



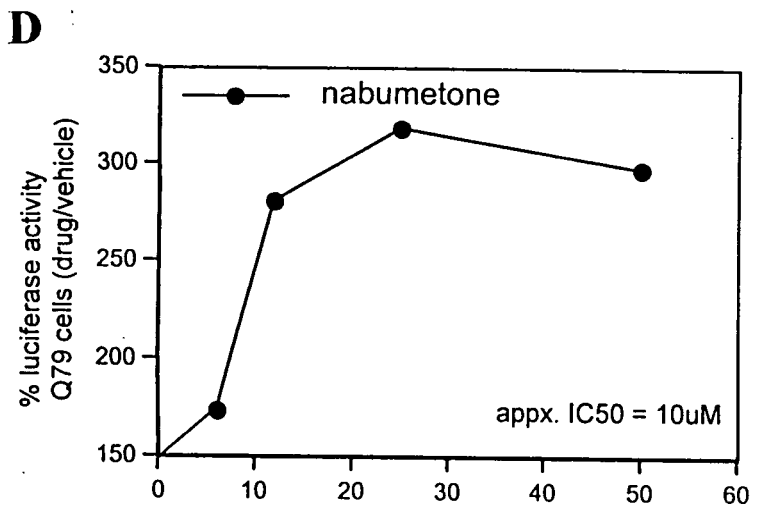
B

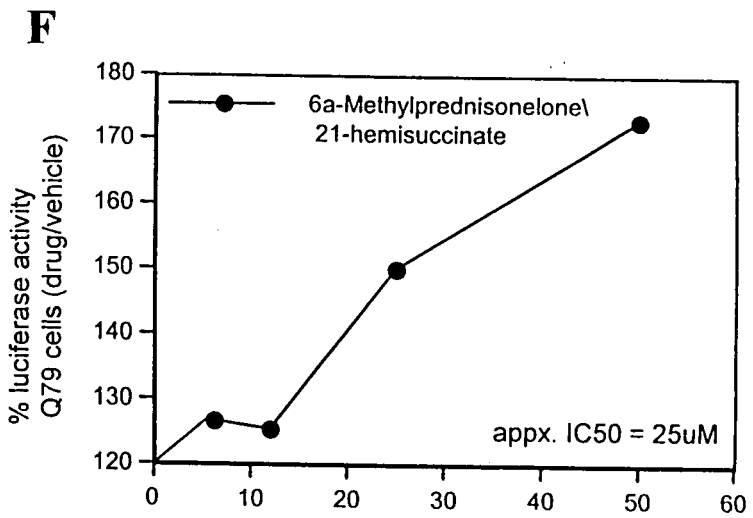


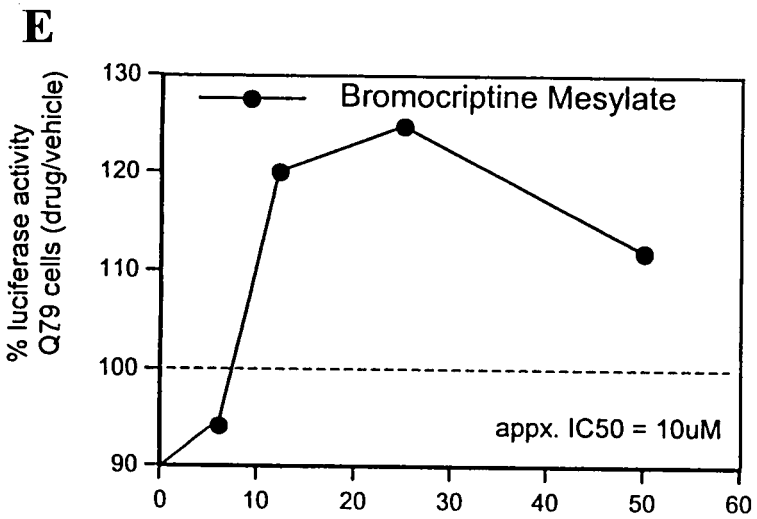
C

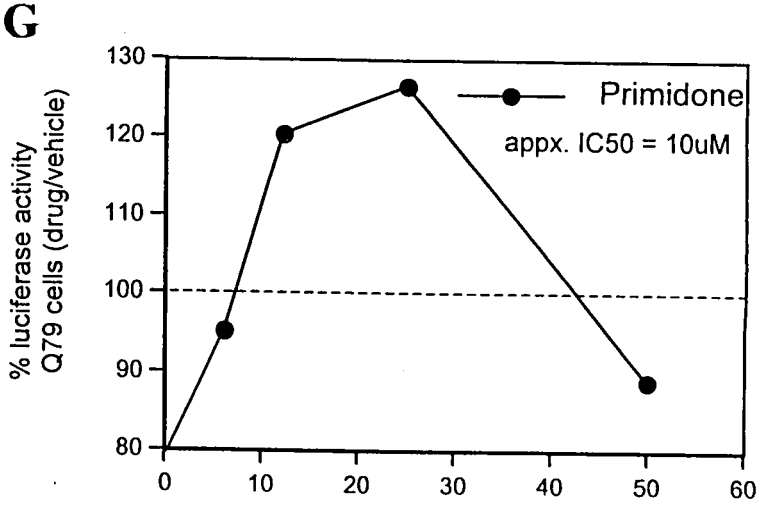


Figs. 17A - 17C

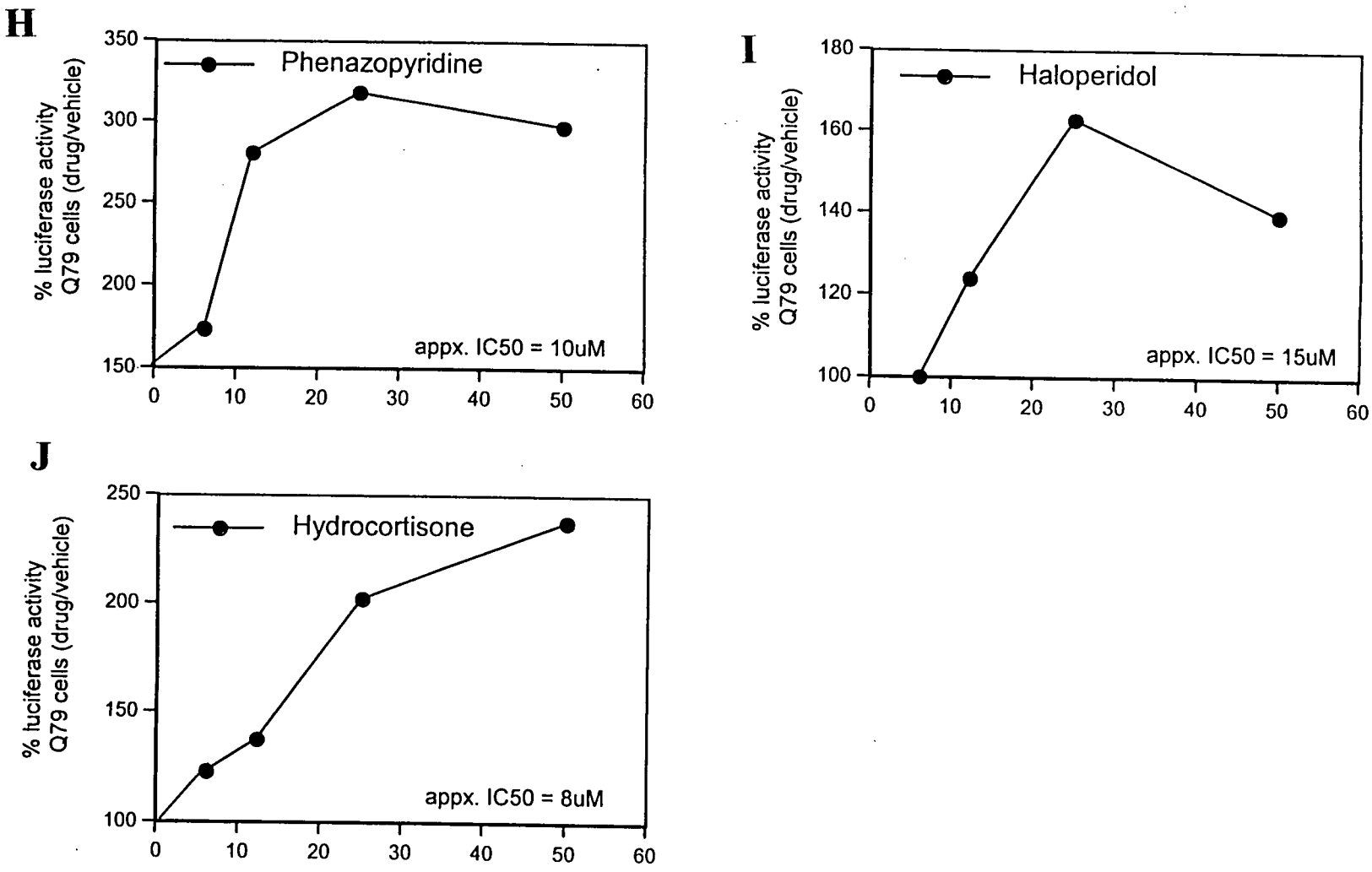








Figs. 17D - 17G



Figs. 17H - 17J

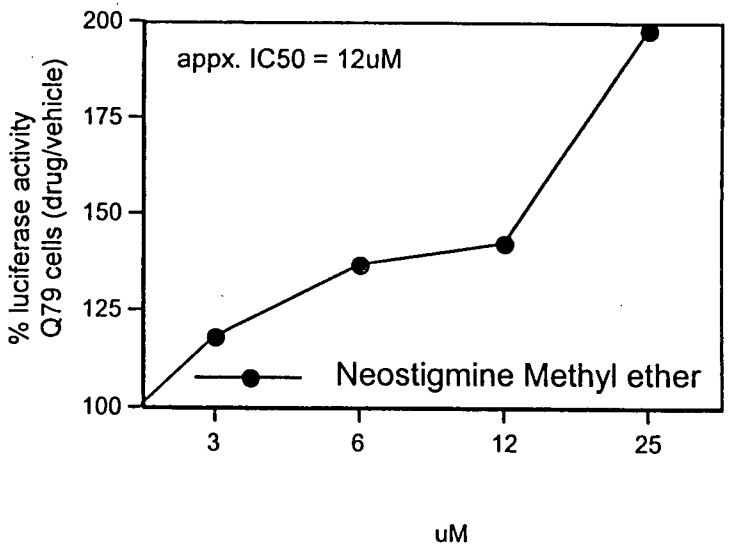
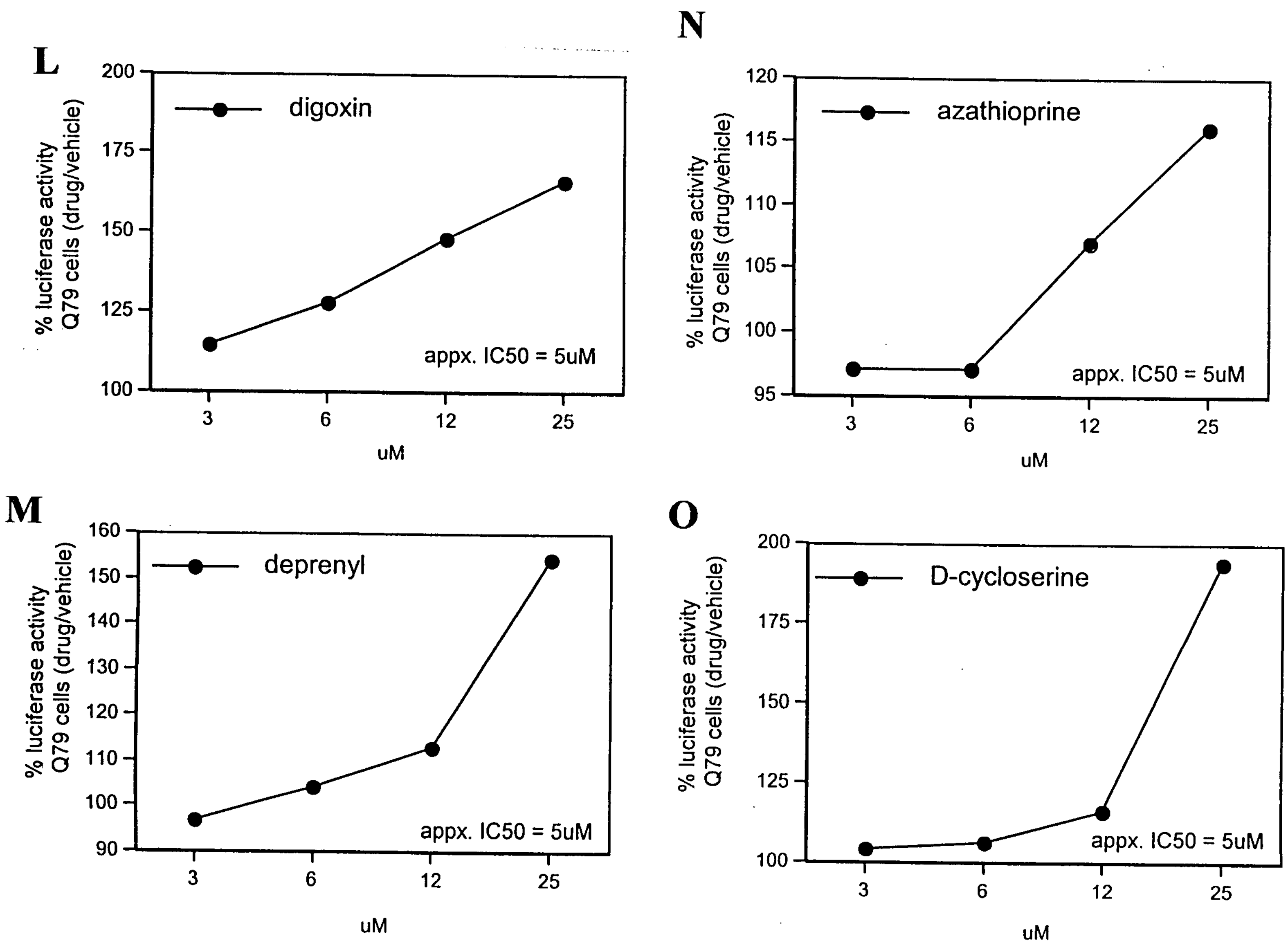
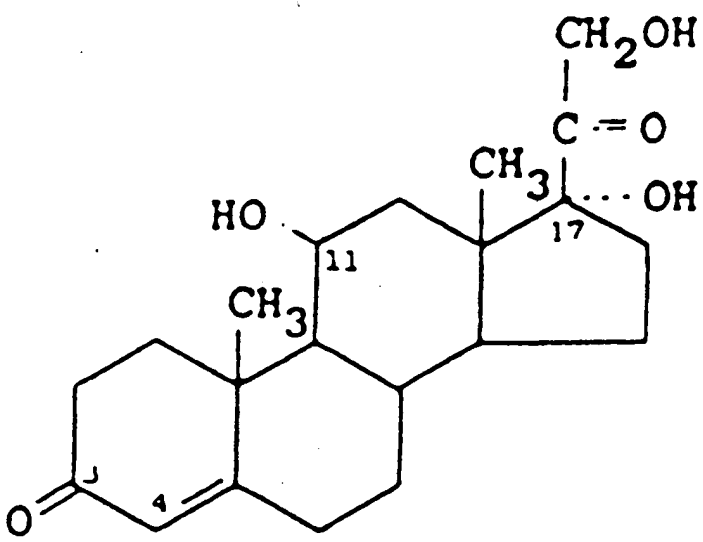


Fig. 17K



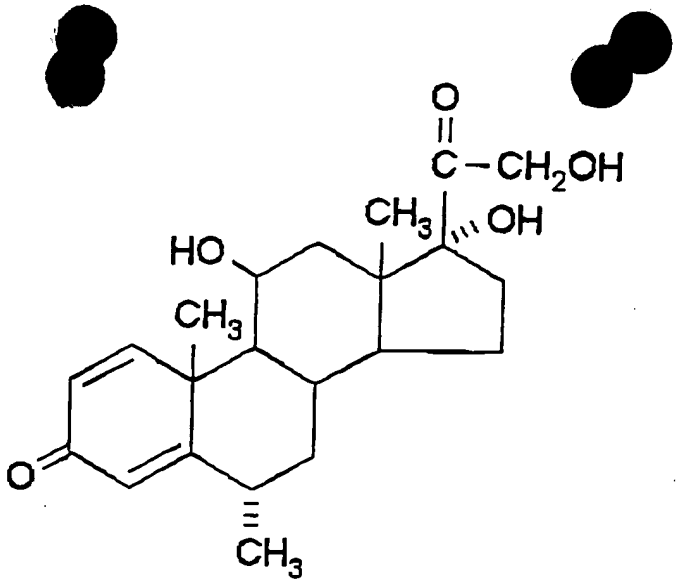
L
% luciferase activity
Q79 cells (drug/vehicle)
200
175
150
125
100
digoxin
appx. IC50 = 5uM
3
6
12
25
uM
N
% luciferase activity
Q79 cells (drug/vehicle)
120
115
110
105
100
95
azathioprine
appx. IC50 = 5uM
3
6
12
25
uM
M
% luciferase activity
Q79 cells (drug/vehicle)
160
150
140
130
120
110
100
90
deprenyl
appx. IC50 = 5uM
3
6
12
25
uM
O
% luciferase activity
Q79 cells (drug/vehicle)
200
175
150
125
100
D-cycloserine
appx. IC50 = 5uM
3
6
12
25
uM

Figs. 17L -17O



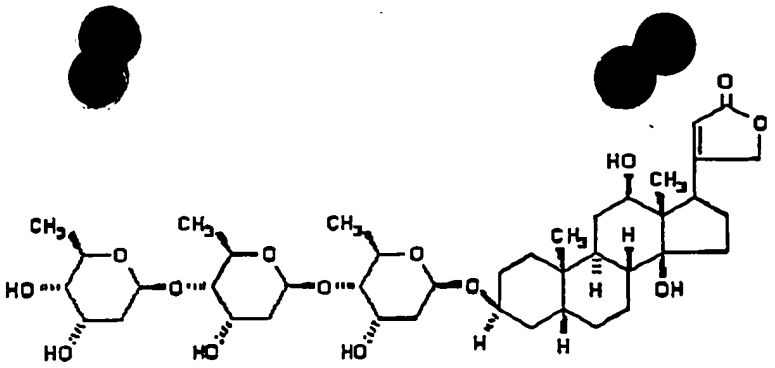
hydrocortisone

Fig. 18A



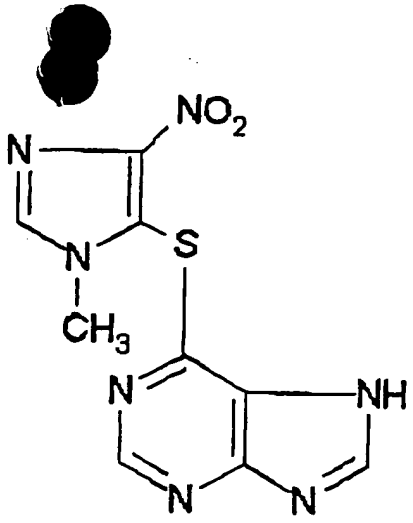
6-∝Methylprednisolone 21-hemisuccinate

Fig. 18B



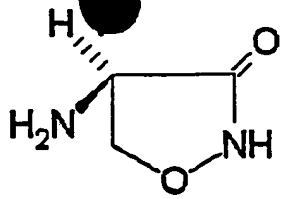
Digoxin

Fig. 18C



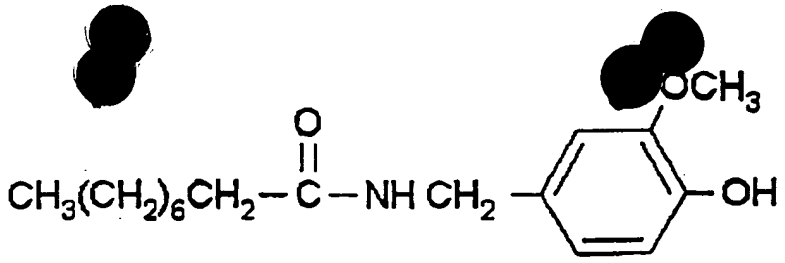
azathioprine

Fig. 18D



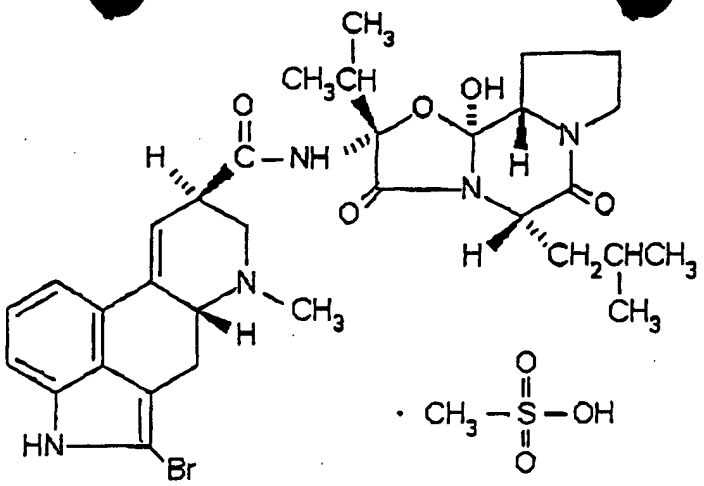
D-cycloserine

Fig. 18E



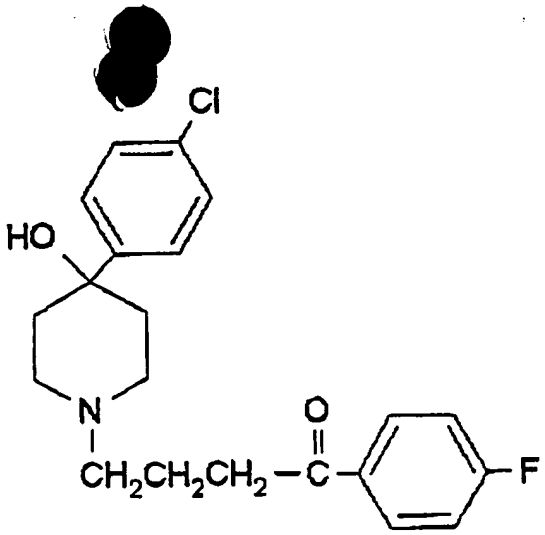
N-Vanillylnonanamide

Fig. 18F



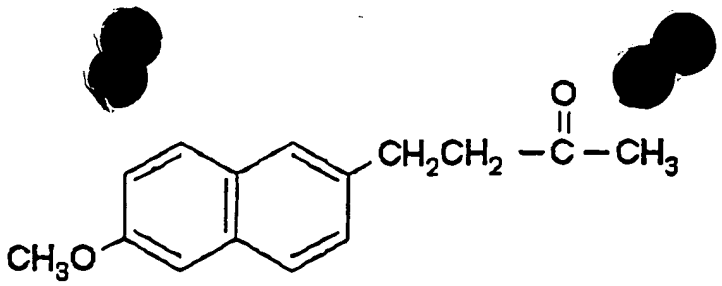
bromocriptine mesylaste

Fig. 18G



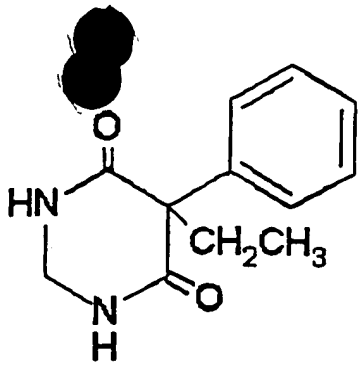
Haloperidol

Fig. 18H



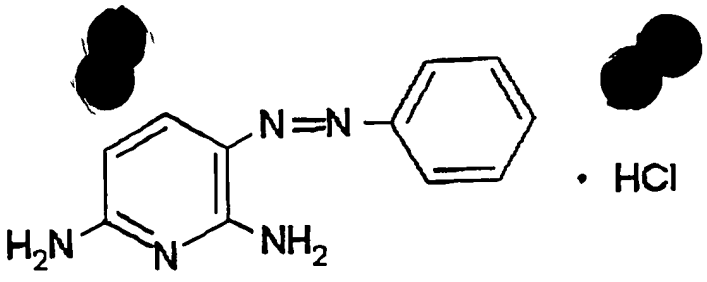
Nabumetone

Fig. 18I



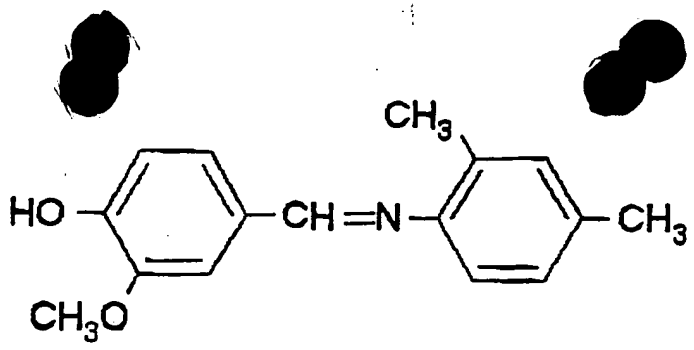
Primidone

Fig. 18J



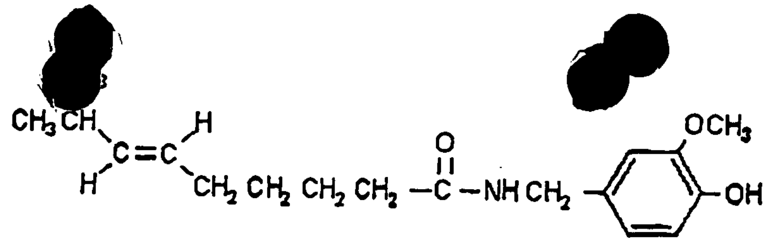
Phenazopyridine

Fig. 18K



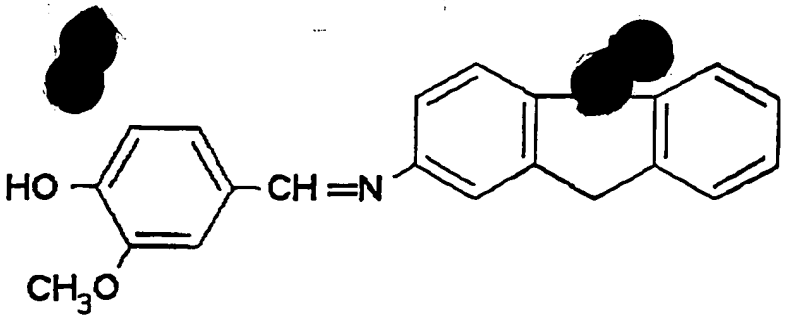
N-Vanillylidene

Fig. 18L



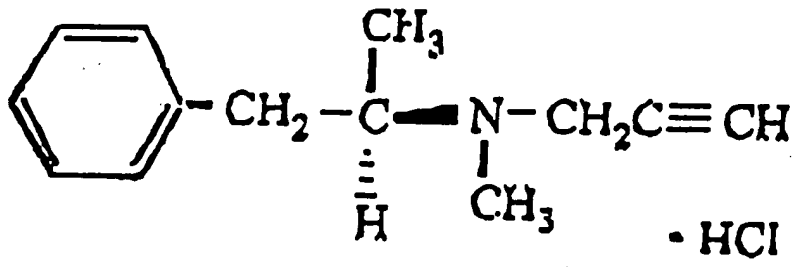
8-methyl-N-vanillyl-6-nonenamide

Fig. 18M



2-(N-Vanillylideneamino)-Fluorene

Fig. 18N



R-(-)-deprenyl hydrochloride

Fig. 18O

Fig. 19